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"Every year about 250,000 American babies are born damaged in some way."
—March of Dimes

Jessyca Russell Gaver

Birth Defects and your Baby

For anxious mothers and all concerned Americans . . . A sane and sensible report on a mounting national problem—its causes, treatment and hoped-for cure.

HERE ARE THE FACTS THAT *MUST* BE FACED—

- 250,000 children are born in the U. S. each year with birth defects. This misfortune strikes about one in every ten American families.
- 20% of all defects are inherited from one or both parents who may not have the defect themselves, but carry it in the genes they pass on to their children.
- The toll in anguish to parents, in medical bills to families, in costs to society for institutional care is incalculable. The defects caused by a single, large-scale epidemic of rubella (German measles) for example, may ultimately cost our country $1,500,000,000.

AND HERE IS THE BOOK THAT HELPS YOU DEAL WITH THEM . . .

BIRTH DEFECTS AND YOUR BABY reports science's latest research into cystic fibrosis, epilepsy, cerebral palsy and scores of other afflictions. It shows how birth-defect problems can be controlled and in some cases even prevented. And perhaps above all, it tells the anxious mother what she can do, even now, to lead her baby to a natural, normal life.

"Let me come in and weep with you,
I who have no little child to die . . ."

—Josephine Dodge Darham

Jessyca Russell Gaver

Birth Defects and your Baby

MAGNUM BOOKS • NEW YORK

A LANCER BOOK

BIRTH DEFECTS AND YOUR BABY

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Dedicated with All My Love to my daughter, Claudia Gaver Grace and her husband, James Walter Grace, Sr. for being the nicest "children" in the world . . .

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Introduction

Until February 26, 1970, when my grandson, James Walter Grace, Jr., was born, I had little knowledge of birth defects. Upon learning that little Jim had an open spine, I sought help and advice for him, and it was given generously by The National Foundation-March of Dimes and two of its grantees, Dr. Louis Cooper of New York University Hospital's Rubella Project and Dr. Alfred Scherzer of New York Hospital's Spina Bifida Project, both in New York City.

Little Jimmy died at three weeks old for two reasons: (1) The pediatrician supposedly taking care of him did not know about the Birth Defects Centers established throughout the United States by The National Foundation-March of Dimes, and, (2) that same pediatrician and his medical associates flatly stated to me that they did not believe handicapped babies should be allowed to live. It was that stark and simple. Not until we threatened a malpractice suit were we allowed, nine days after the baby's birth, to take the infant to the wonderful Birth Defects Center in Newark, New Jersey. But by then it was too late—the open spine had become infected. If little Jimmy had been operated on within 24 hours of his birth, he might have lived.

It was too late to punish the appallingly ignorant pediatrician. All our family could do was to find some way of making certain our baby had not lived—and died—in vain. That was when I got the idea for this book.

Twenty-two other publishers turned it down, so we are indeed very grateful to Irwin Stein, Editor and Co-Publisher of Lancer Books for accepting it, and for the wonderful editing and cooperation of his editor, John Holt. This is the first time that any lay publisher has put out such a comprehensive volume for the families of birth-defect babies.

If this book does nothing more than to help you learn that most birth defects can be prevented, and-or how to live with

your own child's birth defect, then our little Jimmy's brief existence has been truly purposeful.

May God help you, through this book, to recognize that there are already ways of helping these handicapped children. With it, you will find out that Love does, indeed, work miracles!

1

Genetics, Chromosomes and You

If you or a member of your family has a birth-defect baby, you probably feel it is the greatest tragedy of your life. It isn't. It is one of nature's mistakes, but if the baby lives and can be brought home, you've won half the battle. You will find that, apart from his physical or mental problems, he is still a living, breathing bundle of joy much of the time, and that you wouldn't want to lose him under any circumstances.

This book is meant to tell you how you can live more easily with a birth-defect baby, what is available in help and comfort for you, and which organizations and scientific procedures are available every step of the way as your birth-defect baby lives and develops.

Primarily, this book hopes to emphasize that, in many ways, there really is help for your troubled baby and you. But first you must also understand that you don't have to repeat nature's mistake—that science is rapidly developing a number of ways to prevent this happening over and over again in the same family and that this prevention does not always mean having no more children.

It wasn't until the 1960s, however, that the general nonscientific public began to learn there was something called "the genetic code," which is the method by which the forms and functions of all manners of life are transmitted from one generation to the next. The "code" is in the intertwining strands of the basic molecules, deoxyribonucleic acid (DNA) and ribonucleic acid (RNA).

Scientists have learned how to surgically delete "bad" genes in cojoined chromosomes at the time the egg is fertilized and a new life is beginning. This is possible because our bodies are made up of cells, the smallest units of living matter. They fall into specialized groups, the tissues, which are in turn arranged into the heart, liver, brain, and other parts of the body. The cell survives as part of a complex assem-

blage with what is hoped will be a smoothly working organization uniting and controlling all the specialized groups. When something affects this smoothly working organization the result can be one or more birth defects in the baby that is born.

A typical human cell is surrounded by a plasma membrane, which is a living part of the cell, holding its shape, and allowing materials to pass in or out. The cell is divided into two major portions: the nucleus, a small central portion surrounded by nuclear membrane, and the cytoplasm, which is protoplasm containing nutrients and water. The nucleus controls cell division and contains the mechanism that ultimately defines the nature of the cell's chemistry. When the cell is going to divide, a network of granules arranges itself into the series of units called chromosomes, which contain the mast chemical DNA. DNA carries coded instructions for the creation of the companion nucleic acid (RNA) which lines up the specific amino acids required to create vital proteins for the body.

Each cell acts as a machine that does work and converts energy from one form to another. When one or more of its molecular components is missing or functioning improperly, or cannot cope with the strains put upon it, the malfunction often results in what we call a disease. A physical or chemical alteration takes place. Activities of cells change. The result can be arthritis, cancer, or many other ailments, such as birth defects.

Chromosomes are microscopic rod-shaped bodies found in the nucleus of a cell. They contain the genes which carry hereditary factors. Chromosome breakage may occur when certain environmental or chemical influences disrupt the natural conformation of the chromosomes. Breaks are normally found in about one or two per cent of the cells. Breakage in itself is not usually harmful, since cells with broken chromosomes normally die. In rare cases, however, the broken pieces of the chromosome rearrange themselves into unnatural shapes. If this occurs in cells involved with reproduction, it could conceivably happen that the rearrangement of the genes could produce abnormalities in the fetus, which means in the unborn baby.

Chromosome breakage has been shown, in different types of laboratory studies, to be caused by substances in the cells of various plants, and animals, as well as in chemicals. Among

these substances are caffeine, aspirin, LSD, marijuana, mercury pollution, and many others. However, it has not yet been established whether the results of plant, animal, or chemical research are relevant to man's normal use of these substances.

According to the article, "Guide to Human Chromosome Defects," (published in the Birth Defects Original Article Series, Vol. IV, No. 4, Sept. 1968 by the National Foundation-March of Dimes, and written by Audrey Redding, B.A., Associate of the Division of Medical Genetics, Mt. Sinai School of Medicine, New York City, and Kurt Hirschhorn, M.D., Chief of that division) the significance of chromosome breakage is not yet fully understood by scientists. However, from their study of chromosome breakage there has developed the field of cytogenics and recognition of the fact that a cytogenic defect is a variation of the norm in either the structure or the number of chromosomes.

Chromosomes contain genes, which are the units of heredity. The genes are so small that they are nearly invisible. It is many genes together, always in a specific sequence, that make up the structure of a chromosome. Except in a single instance, each cell of the human body has 46 chromosomes. These include 22 pairs of chromosomes which are called autosomes. Each pair of autosomes is different in genetic content, and frequently in appearance, from all other pairs. In addition to these 22 matching pairs, there are two which are labelled X and Y and are called the sex chromosomes. All normal males have one Y chromosome and one X chromosome. The normal female has two X chromosomes and no Y chromosome. The only cells not having 46 chromosomes are the sperm, the ova (egg cells), and the reproductive cells from which they develop. They are considered a single example, however, for the purpose of scientific explanation. When the sperm and ova fuse at conception, they form the beginnings of a new individual who will again have 46 chromosomes in all cells except for the reproductive cells.

The growing human embryo is a biochemical kaleidoscope, its patterns changing from hour to hour. In the first few weeks after conception the chemical pattern of its cells undergo numberless changes, reflecting the release of genetic information inherited from parents, grandparents, and older ancestors back to the beginning of the line of life that led to man. If the genetic information should happen to be

faulty—perhaps because of a mutation in the chemical substance of a gene—a variant chemical pattern may be assembled during chemical differentiation. The embryological cells may then be induced to form a heart with an incomplete septum. Or the arms may be formed so abnormally short that they resemble seal flippers—the congenital malformation known as Phocomelia.

But suppose the genetic information is correct and yet the nine-month infant is born with seal-like flippers for arms? This was the case with the so-called "thalidomide babies," and the example of thalidomide demonstrates the vulnerability of the human being during those kaleidoscopic early days in the womb.

A few molecules of a foreign chemical, perhaps introduced into the embryological circulation from the circulation of the mother, can block or break the characteristic biochemical patterns and cause malformation just as if the genetic information was wrong.

The agents and procedures—foreign chemicals, radiation, viruses, or genes—that are capable of malforming action in the embryo are called teratogens. Teratology, the science of birth defects, is the study of teratogens and their effects in animals and men. It is an important science: congenital (present-at-birth) malformations are a leading killer and disabler of children.

Even so, unless you're a doctor of biological science, you might think of teratology as a post-thalidomide phenomenon. Many laymen first heard of it after the thalidomide tragedy of 1961, when teratologists were called upon to explain how five thousand or more children in Europe had been deformed before birth by some unexplained action of a sleeping drug taken by their mothers.

Teratologists were actually at work for many decades before thalidomide. Beginning with the careful anatomic descriptions of malformations made by some of the early teratologists, the science progressed in the 1930s to biochemical and genetic investigations which may ultimately explain the mechanisms of congenital defects. However, few examples of environmental teratogens had been observed and proven in man in the early days of teratology, perhaps chiefly because before thalidomide teratologists were seldom able to work with large groups of pregnant women exposed to a potential teratogen.

In humans, teratologists are chiefly concerned with serious malformations that require medical attention. A great many congenital malformations do *not* require medical attention. In fact, probably almost everyone carries some variation of body structure which would qualify as "malformation" in anatomy textbooks. Your own malformation may be as noticeable as an extra finger, or as occult (hidden) as an abnormally-placed artery. Such variants from "normal" are often genetically-based and present in the population to greater or lesser degrees. If a majority of persons in a population should come to carry the genes for variant structure (for six fingers, for instance), six-fingered structure could become "normal" and five fingers "malformation." Note, however, that six fingers might become "normal" only because it is a malformation which has no severe effect on the life or health of the person who inherits the genes for variant structure. Genes for fatal or disabling variations are usually self-limiting in a population—the persons who inherit them often die early or do not reproduce efficiently.

If there is an official definition of a congenital malformation, it is probably the one furnished by Josef Warkany, M.D., professor of pediatric research at the Children's Hospital Research Foundation, University of Cincinnati. Dr. Warkany, who began teratological research in the 1930s and is regarded as one of the most authoritative voices in the field, considers a congenital malformation to be "any gross structural change present at birth in a child or an animal." Dr. Warkany includes congenital malformations within a larger grouping of defects called "congenital anomalies." The anomalies include phenylketonuria (PKU), galactosemia, and other biochemical disorders, but these are not considered congenital malformations because they are not anatomic deformities.

According to a Scandinavian medical journal, *Acta Obstet. Gynec. Scand., issue of* 49:299, 1970, a scientist named Furuhjelm expressed the opinion that many infants who have been delivered by cesarean section because of imminent problems are destined to have mental or physical handicaps later in life. Of 112 such infants studied, 23 had scores (in the test given immediately after birth known as the Apgar Score, named for Virginia Apgar of whom you will hear later) of less than 7 at one minute, and at follow-up 25 had mental or physical abnormalities. This would sug-

gest, therefore, that if you can ask your obstetrician to avoid cesarean procedure except in a case of dire emergency to the life of the infant, you might be preventing at least a birth injury, if not a congenital birth defect.

The wide distribution of congenital malformations (about which there will be detailed examples in a special section of this book further along) forces the teratologist to limit his concern to the medically and economically most important among humans, Dr. Warkany explains. Investigators are examining test animals more closely for "hidden" malformations in the hope that they will be equally helpful in determining how abnormal human development is produced.

Teratogenic genes are at least as important as environmental agents as causes of congenital malformation, but little can presently be done to prevent malformations that are due to purely genetic mechanisms. The exceptions are the few genetically determined malformations which can be detected in advance by the science of genetic counselling and amniocentesis. Again, this is discussed fully further along in this book.

Teratogenic genes may contain either faulty or unstable genetic information. Faulty genetic information can cause "mistakes" to be made in organizing the tissues of the embryo; unstable genes may be overridden by, or act in conjunction with, a teratogenic agent or another gene to produce a malformation in the embryo.

Eight environmental agents or procedures, after introduction into the mother are known to be capable of inducing malformations in the embryo. In addition to thalidomide they are: German measles virus (rubella), which infects the mother first, then damages the embryo; the parasite Toxoplasma gondii; ionizing radiation; carbon monoxide; folic acid antagonists (drugs that antagonize an essential nutrient); some synthetic progestins (hormones); and severe iodine deficiency. Many others are suspected, and still more are mentioned as parts of other detailed birth defect examples in a later chapter of this book. One supposition is that the taking of the birth control pill for a length of time may lead to an unknown folic acid deficiency, despite the addition of folic acid to the vitamins given when a woman knows she is pregnant. Lack of folic acid, of course, is a cause of anemia to both the fetus and mother, and anemia itself can be a cause of ingrown birth defects to the growing child.

One problem that naturally comes up is the fact that the most important basic development of the embryo-turning-into-fetus happens in the first 12 weeks of pregnancy when, as a matter of fact, many women don't even know they are pregnant. Both genetic counsellors and teratologists, as well as nutritional experts, mourn the lack of awareness of pregnancy in that crucial time, since bad nutritional patterns and lack of certain vitamins can cause as much damage as the aspirin or other medications the mother may take without recognizing she is pregnant and possibly damaging her developing child.

Teratologists stress the point that most known teratogens have been discovered in the laboratory and have no proven influence on human embryological development. Riboflavin deficiency, for example, has never been shown to affect the human embryo, even though it can be markedly harmful in the rat.

Teratologists are beginning to find some answers to the most basic question facing them: How do teratogenic agents and procedures produce their tragic birth effects? It is known that genes direct growth and development by ordering the placement of essential nutrients (amino acids, vitamins, and so forth) in the right places at the right times. Teratologists believe that teratogenic agents may act by:

(1) Bringing about a deficiency of nutrients, as in riboflavin deficiency. C. W. Asling, M.D., Ph.D., professor of anatomy at the University of California, points out that deficient diets fed to pregnant laboratory rats have enabled scientists to study basic mechanisms of teratogenesis.

(2) Competing with normal chemicals or metabolites for positions in vital biochemical systems, as in the case of certain antimetabolite drugs. Once incorporated into the system, according to James G. Wilson, Ph.D., professor of anatomy at the University of Florida, Gainesville, the competitor renders the system powerless to function normally.

(3) Blocking the formation of ribonucleic acid (RNA), the "messenger" chemical which is one of the first produced in the cell. RNA organizes most of the other intracellular chemicals; suppression of RNA synthesis by such drugs as the antibiotic actinomycin D interferes with cell differentiation at the earliest point.

(4) Destroying cellular components, as in the case of certain viruses, ionizing radiation, and some toxins.

Other mechanisms have been suspected, and still more are waiting to be discovered. Science is only now beginning to realize the high frequency with which some infections among the many viruses, bacteria, protozoa, and other infectious agents in the environment do occur in pregnant women and the spectrum of birth defect damage that they can cause. Because infectious diseases are often detectable, preventable or treatable by the use of appropriate techniques, expanded efforts for the detection and prevention of them are constantly needed.

John L. Sever, M.D., Ph.D., chief of the Section of Infectious Diseases at the Perinatal Research Branch of the National Institute of Neurological Disease and Stroke, spoke at the Symposium on Birth Defects of the National Foundation-March of Dimes, held on January 27, 1971, and said that to recognize infection during pregnancy or in the child there must be used any one or all of the following techniques: (1) clinical recognition of a disease; (2) demonstration of the micro-organism, or (3) development of increased immunoglobulins and specific antibodies. Date for the 60,000 women who participated in a collaborative perinatal research study showed that more than five per cent of the women reported clinical viral infection during pregnancy. Specific infections such as mumps, rubella, chicken pox, virazoster, viral bronchitis, measles, viral conjunctivitis, viral pneumonitis, and infectious mononucleosis occurred at rates of between one and 15 per 10,000 pregnancies.

However, good news from the National Institutes of Health (NIH) in April, 1971, stated that pregnant women who are susceptible to rubella can be identified through blood tests, then vaccinated in the first few days after delivery. Women of child-bearing age are the most important persons to protect against rubella because of the disease's frequently disastrous effects on the fetus, but the possibility of administering the vaccine during the early weeks of an unsuspected pregnancy has limited its use to girls past puberty. The simplicity, speed, sensitivity, and reliability of this new test permits rapid and effective evaluation of immune status. Two to four days after childbirth, 74 of the rubella-susceptible patients in the study received what is called the Cendehill rubella vaccine. Sixty-eight patients developed sig-

nificant levels of rubella antibody. Only a few patients experienced mild transient reactions to the vaccine. Drs. D. M. Horstmann, H. Liebhaber, and E. I. Iohorn, who conducted this study at the Yale University School of Medicine, suggested that rubella vaccine administered in the immediate post-partum period is both safe and effective, but that physicians should determine whether a woman is susceptible to rubella as a routine part of prenatal care. Vaccination after the birth of the first child should considerably reduce the number of women susceptible to the virus, and thus reduce the rubella problem, along with keeping up the school and health department programs of vaccinating younger schoolchildren who might get rubella and infect pregnant women.

However, it is just as important to consider the factors determining the teratogenicity of a drug or other environmental agents surrounding the pregnant woman. These are (a) the time at which it affects the developing human being and (b) the dosage given. As mentioned earlier, the time of greatest damage to the fetus is during the embryo stage of development, those first few weeks after conception when the process of chemical differentiation is at its peak. However, the human fetus can also suffer brain and other organ damage from certain infections when it is already several months old in development. Mental retardation is linked with severe malnutrition occurring either in the mother or child during the early stages of development (although it can also come through viral infections and biochemical problems during the embryonic stage). Malnutrition or malnourishment during fetal cell division may produce a permanent deficit of as high as 15 per cent in the number of brain cells. If the nutritional status is below normal, both before and after birth, the brain cell deficit may be as high as 60 per cent.

Just how early malformation may be induced in the human embryo is not definitely known in all instances. It is probably so early that only those women knowing they are trying to become pregnant could avoid swallowing or doing anything that might have an adverse effect on the newly-developing embryo. Since all commonly malformed human organs are in the process of formation by the twentieth to sixtieth day after conception, the twentieth post-conception day might be the earliest date at which the human embryo could be malformed. But few teratologists would want to set a date so firmly on present evidence.

One frequently ignored possible factor in birth defects could be the matter of vaginal douching. It is well known that many reckless young women allow their husbands to push them out of bed after intercourse, to run into the bathroom for a fast douche, instead of using other birth control procedures. Anyone who understands the psychology of the female knows this is usually one whose husband doesn't want her to get pregnant, but neither does he want to be bothered with having to use a condom, or get involved with the waiting for his wife to prepare the jelly for her diaphragm. He never stops to consider that it may be just as damaging mentally and emotionally to his wife to be pushed out of bed immediately after intercourse, in order to take that fast douche, as it is important to him physically and emotionally to have copulation with her. The wife who allows herself to be pushed out of bed for a douche is not in the least interested in birth control but is going along with the douching routine so her husband won't "blame" her when she becomes pregnant. What she does not realize is that fewer and fewer doctors believe in douching even as a routine cleanliness measure, let alone as a birth control factor. The natural secretions of the vagina are intended to kill any stray bacteria. If you get a discharge or other vaginal problem, your doctor will prescribe something to use, if he knows you honestly do not want to get pregnant. Say you do douche frequently, however, and are unwittingly pregnant, how do you know you may not unwittingly be upsetting the chemical balance of your newly-created embryo, which might then contribute to an eventual birth defect? You don't know, and neither do the experts—yet—but it's something to be careful about.

However, as a rule, congenital malformations of the fetus cannot be induced after an organ has completely developed. But cleft palate is an example of a very serious congenital birth defect to be found during the stage of chemical differentiation. The human palate usually closes during the eighth post-conception week. If palate closure fails to take place for any reason, the congenital malformation known as cleft palate results. This does not mean, though, that a teratogenic agent or gene cannot act earlier than the eighth week—say at the sixth week—to disrupt some biochemical system that is essential to the process of palate closure. The same might be said for spina bifida (open spine, which ordinarily closes during the 12th post-conception week. It is not

known what causes it to remain open in cases of this malformation, but something does, and science is still trying to figure it out.

Experts say that the place to look for drug teratogenicity in man is in man and have proposed that a registry be set up to keep a record of all drugs administered to pregnant women. Doctors and hospitals would have to agree to report to the registry. Whenever a malformed baby is born to a woman who has received drugs, the record would show what drugs the mother had received, at what time, and in what dosage. Also in the registry would be a record of the total number of pregnant women who had received the same drug at a time critical for the embryo; thus investigators would also know how many times the drug was *not* associated with a malformation—information which is lacking for most drugs. Science investigators would be furnished with much-needed statistical information which they now frequently lack. Suppose, for example, that during one year a certain drug is given to 100 women in early pregnancy. Then further suppose that five of the babies born to these women have cleft palets. Today, there is no way for scientific people interested in birth defects to be informed of the five cleft palet cases, or for them to learn essential facts about what drugs are given to each mother. Most important, even if they learned of the five cases they could not determine the statistical relationship between drug and malformation. If 10,000 pregnant women had received the drug in one year, for example, five cases of cleft palet due to teratogenic genes might naturally have occurred among their children, anyway. But five cases of cleft palet in 100 births would be a statistical incidence too high to explain as "natural occurrence." If teratologists had a way to discover the five-in-100 ratio, they would have some strong evidence indicating teratogenicity of this particular drug in humans.

Should drugs be given to a woman in early pregnancy? This is a question that must be resolved by the pregnant woman's physician in each individual case, Dr. Monie says. If the drug is one that is suspect, the possibility of its damaging the embryo must be carefully weighed against the likelihood of death or serious disability in the mother if the drug is withheld. Definitely, the pregnant woman should take *no* drugs—not even an aspirin—unless they are prescribed by her doctor, teratologists warn. Taking unprescribed medicines

could be potentially dangerous for the embryo, especially if other drugs are being taken.

"We are just beginning to understand something about the teratogenic potential of drug combinations," is another explanation. "It is possible that two or more drugs which are subteratogenic when each is taken alone may become teratogenic when combined accidentally. In some cases, the drugs may so enforce one another's actions that their combined effect is greater than the simple summation of individual effects."

Actually, genetic engineering—the manipulation of material in living cells so as to remedy genetic defects—has been brought much closer to reality by a group of scientists at Oxford University, England, according to a story published in the *Manchester Guardian* in March, 1971. In a series of experiments, Professor Henry Harris and his assistants inserted into a deficient mouse cell the specific genetic material needed to enable it to function properly. In this case, the deficiency was the inability to produce an enzyme called inosinic acid pyrophosphorylase (IAP), a genetic fault which in humans leads to a rare and fatal condition known as the Lesch-Nyhan Syndrome. From the point of view of ultimate clinical potential—which Professor Harris says is considerable, but still at some distance—the "most encouraging" aspect of the success is that the fragments of genetic material have been transferred to mouse cells without the receiving cells becoming in any way "foreign" to the mouse from which they came. This means that it might be possible to remove selected cells from an animal (or human) with some genetic deficiency, manipulate them in the laboratory, grow the corrected cells into a larger colony, and then re-inject them without problems of the kind that have rendered transplants uncertain. Naturally, the fate of such re-injected cells is not yet predictable, but it is possible that they would be incorporated into the body structure and ultimately dominate the formerly deficient functions. Subsequent series of experiments at Oxford will, in fact, aim at repairing the genetic defect in an IAP deficient mouse. Parallel experiments will be carried out to confirm the belief that the technique of transfer operates with human and other animal cells, as well as those of the mouse. The group already knows that it works with other cell systems, but confirmatory experiments with specific deficiencies are an essential part of the progress, necessarily slow, toward clinical

evaluation. The transfer was disclosed in an issue of the journal *Nature New Biology*, published in England. It takes place during the processes which follow the fusion and continued growth of cells from different species of animals. In the successful experiment normal red blood cells from a chicken were fused with genetically deficient mouse cells. In this situation there exists a single cell envelope, but two nuclei, which, because they are from different species, have growth rates which are "out of phase." One nucleus therefore starts to divide before the other and, when this happens, the second nucleus (in this case the chicken cell nucleus) is rapidly broken down in a process called "chromosome pulverization."

We must repeat that chromosomes contain all the genetic information of an organism. The Oxford group reasoned that, since at the time of division the deficient nucleus is without an enclosing membrane, it was possible that information fragments from the pulverized material might fit into the defective regions of the nucleus' genetic material. The structural similarities are known to be great, and therefore it was hoped that the genetic fragment from the chicken cell, with produced IAP, would, with some kind of chemical lock and key, fit into the deficient cell's material. By growing the cells in an environment in which only those that had been repaired could survive they demonstrated that this was the case. Examination of the repaired genetic material showed that the repair was invisible. Further, the cells had apparently incorporated only the material needed for repair, and none of the other characteristics which would lead to problems of immune reaction. Similar transfers have since been carried out from toad to mouse cells and mouse to hamster cells, so the outlook is indeed bright.

Only in the last year have research findings been announced, at a meeting of the American Society of Clinical Investigation in Atlantic City, New Jersey, that involve basic genetic mechanisms in cells of the highest forms of life—the mammals, including man—thanks to a team of scientists from the National Institutes of Health. The NIH scientists reported achieving the first test-tube isolation of certain chemical factors that spark life in mammalian cells, and they incorporated these factors into a cell-free brew that enabled them to study systematically individual mammalian biochemical life processes, for the first time, outside of living cells. In this way the NIH scientists can study fine points in the synthesis

of one of life's most vital constituents—protein—and will be able to biochemically "dissetic" human genetic diseases, one by one. These include hereditary afflictions ranging from diabetes and schizophrenia to cystic fibrosis, sickle cell anemia, and a host of other blood diseases, plus various neurological forms of deafness and blindness, many now incurable, but all disorders that are commanding a growing amount of the country's medical concern and service.

Naturally, hereditary disorders are assuming a larger place in medical practice today and this can account in part to the gradual decline in childhood diseases and deaths due to malnutrition and infection.

During the 1971 proceedings of the American College of Physicians, Dr. Curt Stern, emeritus professor of zoology at the University of California, Berkeley, declared that at least 25 percent of the hospital beds and other institutional facilities are believed to be occupied by persons with physical or mental illness or defects, related in full or in part to genetic control. According to the report in the May 17, 1971 *Modern Medicine*, Dr. Stern suggested that the future role for genetics may extend to genetic engineering, since the formative stage has led to the recognition of genes as decisive agents in health and disease, and genes are linked to their effects by chains of reactions which can be controlled or altered by therapeutic means. For example, he pointed out, rational dietary treatment is successful in some genetic diseases while surgery is effective in others. He also stated that although many genetic diseases cannot yet be treated, some control is becoming available for an increasing number. In addition, early prenatal diagnosis of abnormal metabolism or chromosomal makeup is now possible (see our sections that follow on amniocentesis and genetic counselling), as well as through chromosome study.

Modern Medicine's story also revealed that Johns Hopkins University in Baltimore, Maryland has listed and catalogued genetic disorders in three areas—autosomal dominant, autosomal recessive, and X-linked disorders. Dr. Victor A. McKusick, head of the school division of medical genetics, said that in 1970 the list grew to 396 autosomal dominant, 353 autosomal recessive, and 79 X-linked disorders—a total of 828. Also, 1003 other genetic disorders were listed as tentative, waiting to be proved, or discarded.

Abnormalities of the chromosomes have been related to six

categories of disease, all congenital: abnormalities of sexual development, mental retardation, complex malformation syndromes, 20 percent of spontaneous abortions, acquired abnormalities in neoplastic disease, and behaviorial abnormalities.

Among suggestions made by Dr. McKusick concerning genetic counselling is that the total cost of a genetic disorder should be measured in both recurrence risk and the burden of the particular condition. For example, color blindness has a high recurrence risk, but is a low burden. He listed as potential treatments for genetic disease: replacement of the gene product, vitamins for "vitamin-dependent" inborn errors of metabolism, elimination from the body, enzyme induction, and specific dietary restrictions.

Genetic Counselling:

The best thesis we have seen or heard on genetic counselling for parents and prospective parents was given at the June, 1970, American Medical Association Convention by Hymie Gordon, M.D., F.R.C.P.E., Department of Medical Genetics, Mayo Clinic, Rochester, Minnesota. We give you here part of what was given to members of the press there:

"In most medical practices, advice about the possibility of the recurrence of a genetic disease or a congenital anomaly is usually sought by parents who have already produced an affected child, or by prospective parents, one of whom may be affected by or has a family history of what may be a genetically-determined disease. Such advice is requested not only in respect of the very rare genetic disorders, but also in respect of fairly common conditions such as epilepsy and diabetes. Given a precise diagnosis, a carefully-elicited family history and an adequate understanding of the principles involved, the genetic prognosis can be given in most cases as a probability statement with a high degree of confidence. But such a statistical statement is not enough. As in the case of any other medical consultation, discussion of the implications of a genetic prognosis must be related to the personal circumstances of the individuals concerned and requires a high degree of personal doctor-patient rapport. Where the genetic counsellor himself is not in a position to establish this kind of rapport, his findings should be transmitted and interpreted to

the parents or prospective parents by their regular family doctor.

"In the circumstances of family practice, there are two relatively common situations in which genetic counselling may be sought. The most common situation is that which arises when a child is born with an inherited disease or with a congenital anomaly; the parents, naturally, will want to know if the condition is likely to recur in their subsequent children. The other situation arises premaritally, when a young couple are considering marriage and one of them has a relative with what might be a heritable disease: they want to know what are the potential risks of this disease affecting their children . . .

"It is only in recent years that requests for genetic counselling have begun to be an important part of medical practice. Two coincidental historical processes, operating in advanced societies, are mainly responsible. . . .

". . . diseases in which genetic factors predominate have become relatively more numerous and demanding of attention. . . . During this half-century (from 1916 to 1965) the total infant mortality rate has fallen from 101 to 25 per 1,000 live births; the infant mortality rate for the diarrheal diseases (enteritis, colitis, dysentery, etc.) has fallen from 24.1 to 0.6 per 1,000 live births, that is to one four-hundredth of the earlier rate. However, the infant mortality rate attributed to congenital malformations in 1965 still remains at a level of more than one-half of the 1916 rate (6.8 and 3.6 per 1,000 live born in 1916 and 1965 respectively). . . . Since 1940, a greater proportion of infant deaths has been caused by congenital malformations. . . . There is also a considerable absolute frequency of the genetically-determined diseases encountered in medical practice. As an absolute benchmark against which to compare specific risks in affected families, most genetic counsellors use the estimate that 2.5 per cent of all live-born children will manifest a major heritable or developmental disorder at birth or soon after. There is reason to believe that this is an underestimate and if one includes those inherited or partly-inherited traits which become manifest only in later life, the frequency approaches five per cent.

"The second historical process, which has almost coincided with the epidemiological one, is the extraordinarily rapid development of scientific understanding of the principles of ge-

netics. . . . Today genetics is securely established as one of the most fundamental biological sciences. . . . Most young people are well aware of the possible implications of hereditary diseases in their families and when they seek genetic counsel they expect a rational explanation of their problem and their genetic prognosis. Unfortunately, even in this year and in this sophisticated society one still hears of practitioners wrongly reassuring parents with an affected child that 'lightning never strikes twice in the same place,' or giving the opposite but equally erroneous advice that because it is an inherited disease 'all your children will be abnormal'. . . .

"When confronted with a trait or disease affecting several members of the same family, the question arises: 'Is the trait or disease mainly the effect of the peculiar genetic endowment of the family or is it mainly owing to common factors in their environment?' Sometimes there is no difficulty in answering the question. In the Hapsburg Royal Family of Europe, from the 15th century onward, many members in successive generations have had a protruding lower lip and an unusually shaped mandible; there is little doubt that this is an inherited characteristic. In a family I recently studied in Cape Town, South Africa, the mother and four of her seven children had a narrow pointed skull, protruding eyes, a parrot-like nose and maxillary hypoplasia (Crouzon's syndrome of craniofacial dysostosis); one has no difficulty in accepting this syndrome as genetically-determined. But when one is consulted by the normal parents of three children, two of whom have cleft lips, what does one say then about the relative roles of heredity and environment . . . ?

". . . as one moves across the spectrum, one encounters the common birth defects such as cleft lip, clubfoot, and the neuraltube anomalies; complex genetic factors interact with environmental factors to produce them. Several conditions which develop in later life—diabetes, hypertension, rheumatic fever, asthma, and others—may also be found in the central part of the spectrum as examples of disease of multifactorial causation. Further along the spectrum, moving towards the 'environmental end,' one encounters the cancers; with rare exceptions, environmental factors predominate in their etiology, but there is evidence of some degree of genetic susceptibility in most of them. The infectious diseases lie very near the 'environmental end' but even here there is

some evidence of inherent differences in susceptibility. At the extreme 'environmental end' of the spectrum one can place the effects of injury.

"The diseases at the 'hereditary end' of the spectrum and the multifactorial diseases in the center are the ones with which genetic counselling is most concerned. . . . With a few exceptions, the diseases at the 'hereditary end' of the spectrum are mostly very rare. The exceptions include sickle-cell anemia, glucose-6-phosphate dehydrogenase deficiency, and hemolytic diseases of the newborn associated with rhesus blood group incompatibility. Most of the others have frequencies of much less than 1-in-1000 live births. But although they are individually rare, there are an appalling number of them. McKusick has prepared a catalog of these 'mendelian' conditions which includes no fewer than 1,545 entities, the diagnostic pattern being well established in 692 of them. . . .

"Once the diagnosis and the pattern of inheritance are established, the genetic risks can easily be stated (by the genetic counselior) as far as future children are concerned. For diseases showing the autosomal-dominant pattern the risk to future children is 1-in-2; for autosomal recessives it is 1-in-4, and for X-linked recessives it is 1-in-2 for subsequent sons or 1-in-4 for all subsequent children. But the task of the genetic counsellor does not stop there because, especially in the case of recessive traits, other relatives will want to know if they are carriers of the mutant gene. Again, the risks can be stated quite easily: for example, 2-in-3 of the siblings of a person affected with a disease showing the autosomal-recessive inheritance pattern will be carriers, and half of the sisters of a man with an X-linked disease will be carriers. However, in an increasing number of conditions, it is possible to identify the carriers with even greater precision: often with 80 to 100 per cent confidence.

"Sometimes it is just a simple matter of clinical examination. . . . More often biochemical studies are required which may be simple or complicated. . . . In several of the neurolipidoses (Tay Sach's disease, Batten's disease, etc.), the carrier state can often be detected by the presence in the peripheral blood of abnormal lympocytes with vacuolated cytoplasm. It is much more difficult to detect the carriers of galactosemia in which the enzyme galactose-1-P-uridyl

transferase is lacking. Most carriers have reduced levels of enzyme activity but the range is wide and there may be overlapping with the normal range. Carrier detection is most valuable in the case of the X-linked diseases: the sisters and the maternal aunts of affected boys are most anxious to know if they are liable to produce affected sons, and they can often be advised (by the genetic counsellor) with a fair degree of confidence. . . .

"Sometimes a child is born with a disease known to be determined by an X-linked mutant or by an autosomal mutant with dominant effect (e.g. achonodroplasis) but there had never been a case of this disease in the family before. Such a sporadic case is often owing to the development of a fresh mutant gene in the gonad of one of the parents as a result of the action of a mutagenic agent. If such a fresh mutation develops in a cell of the father's germinal epithelium, the clone of spermatogenic cells derived from that original mutant-carrying cell will all carry the mutant gene and it will appear in the spermatozoa derived from these spermatogenic cells. If one of the mutant-bearing spermatozoa fertilizes a maternal egg, the child will be affected. The risk of the disease recurring in subsequent children will depend on the proportion of the father's spermatogenic cells carrying the mutant. If the mutation occurred at an early stage in the development of the father's germinal epithelium, a relatively large proportion of his spermatogenic cells will be affected, and there will be a greater risk of the disease recurring in his children.

"If the mutation occurred later in the development of the father's germinal epithelium, a relatively smaller proportion of his spermatogenic cells will carry the mutant gene, with a lesser risk of the disease recurring in his children. Unfortunately, there is no way of telling the extent of the mutation in the father's germinal epithelium so it is not possible to give a precise estimate of the genetic risk in subsequent children. Most of the available evidence, however, indicates that the recurrence risk in siblings is quite low and is of the order of about ten per cent. The risk of recurrence in the descendants of the affected person will be the same as in the case of any other mutant gene which is X-linked or shows the autosomal-recessive pattern of inheritance.

"Parental consanguinity (meaning: to have the same ancestry, to be related by blood) is an important factor in the case

of rare disease determined by autosomal mutant genes with recessive effect. Furthermore, when large scale population surveys are carried out it is found that the frequency of children with congenital anomalies is three to four times greater when the parents are consanguineous than when the parents are unrelated. However, at the individual level, the risks are very small and marriage is not necessarily contraindicated, provided that one can be reasonably sure that there are no relatives with genetic diseases or congenital anomalies. Showing the consequences of an ill-advised marriage between first cousins-once-removed, the wife had two nieces with Hirschsprung's disease; they were also first cousins-twice-removed to her husband. The husband and wife had one-sixteenth of their genes in common and it is not very surprising that they also produced a child with Hirschsprung's disease.

"Regarding the proportion of genes in common between persons of different degrees of relationship . . . for example, cousins have one-eighth of their genes in common. This means that if a man carries a rare mutant gene with recessive effect, there is a probability of 1-in-1 that his first cousin will carry the same gene. No matter how rare that gene is in the general population—it can have a frequency of one-in-a-million—if a carrier marries his first cousin, there is a 1-in-8 chance that she will also be a carrier of that gene. . . .

"In most cases of the common congenital anomalies, the risks of recurrence in subsequent siblings after the birth of one affected child are relatively small—usually less than five per cent. Among the commonest congenital anomalies are those affecting the central nervous system: anencephaly, spina bifida, and hydrocephalus. The available data concerning the recurrence risks for this group of anomalies (show) that the risk to a subsequent child after the birth of one affected child is five per cent; after the birth of two affected children, the risk to subsequent children increases to 12 per cent.

"In . . . pyloric stenosis . . . the sex differences are noteworthy. Pyloric stenosis is much more common in males than in females, but if a female child is affected the recurrence risk is much greater than when a male child is affected. In fact, a woman who has had pyloric stenosis has an 18 per cent chance of producing a similarly affected child. This risk, though quite high, will probably not deter her from having children, but it will alert her physician to the correct

diagnosis if one of her children does begin to vomit in early infancy. . . .

"Occasionally, even one of the most common congenital anomalies with low recurrence risks can spring a surprise. For example, in rare instances, cleft lip and or palate may recur in several members of a family with the pedigree showing the autosomal-dominant inheritance pattern. The clue which should alert the genetic counsellor in these cases is the presence of two small pits in the lower lip. When these are present, the risk that the cleft lip and-or palate, will recur goes up abruptly from less than five per cent to nearly 50 per cent. . . .

"*Precise Diagnosis.* Before one can even begin to consider the genetic prognosis, the disease in question must be accurately identified. A vague history of someone in the family being mentally disordered is not enough. Even a diagnosis such as 'muscular dystrophy' or 'ichthyosis' is inadequate: there are several clinical variants of each of these and the responsible gene mutations may be autosomal with dominant or recessive effect, or they may be X-linked. It is also essential to be certain that one is dealing with a primarily genetic disorder and not with a clinically similar acquired disease, a so-called 'phenocopy.' For example, porphyria variegata is the effect of a single autosomal gene mutation but its phenocopy, prophyria cutanea tarda, is usually acquired with genetic factors playing just a subordinate role. This kind of problem often arises in the case of deafness: not only are there many genetically-determined kinds of deafness, but there are also many acquired varieties. For obvious social reasons, deaf men and deaf women are frequently attracted to each other and when marriage is contemplated, there is often the fear of deafness in the children. Careful attention to diagnostic detail often reveals that each prospective parent is homozygous for a different pair of mutant genes, or that one or both of them has an acquired form of deafness. In either case, the risks of producing genetically deaf children are minimal. Of course, if either parent is deaf because of an autosomal mutant gene with *dominant* effect, there is a 1-in-2 risk of deafness in each child born to them.

"Incidentally, the fundamental importance of diagnostic precision emphasizes the importance of the physician in genetic counselling. For many years, before the medical profession awoke to its importance, genetic counselling was usually

left to the local zoologist, anthropologist, or other biologist. These scientists performed a most useful function, for which the medical profession should always be grateful, but they worked under several handicaps, particularly in respect of diagnosis. The first step which the medically qualified genetic counsellor takes when he begins a case-study, is to verify the diagnosis. Because of the individual rarity of many of the inherited diseases, the medical genetic counsellor often has the advantage over even his specialist colleagues in having had more experience with these conditions. In my own practice, it is more common for patients to be referred by my colleagues for primarily verification of the diagnosis and only secondarily for counselling, than for genetic counselling alone. Without disparaging the contributions of the biologists, I am convinced that genetic counselling is part of the art of medicine and is most properly performed by physicians.

"*Adequate family history*. In most instances, the family history need only be elicited in respect of the first three degrees of relationship (i.e. to include parents, siblings, and children; grandparents, uncles, and aunts; and first cousins). Despite its infrequency, an inquiry into possible parental blood relationship should not be overlooked. It is beyond the scope of this outline to discuss the technique of eliciting the family history but I must stress the importance of assessing the reliability of one's informant, and of checking the information about other possibly affected relatives. This may be done by obtaining a report from that relative's doctor, or a hospital report, or a death certificate. The family doctor can make an important contribution; often he knows more about the families' medical history than do individual family members; and when there is confidential family information to be imparted he is often in a better position to be the confidant than an unfamiliar consultant. . . .

"In counselling, I try to present the facts of the case as accurately as possible, and to answer the questions put to me as clearly as I can. Often, the parents or prospective parents return for a second consultation after having discussed the results of the first consultation at home. People who are intelligent enough to seek genetic information are usually intelligent enough to understand it; hence I only present the relevant facts and I do not try to persuade the parents to have babies or not to, nor do I advise young couples to

marry or not to: I supply the facts, but they are left to make up their own minds.

"But I must admit that I prefer to take the optimistic view whenever possible. If young parents have produced an anencephalic baby as their first child, the risk of recurrence is only about five per cent; but when the parents are terrified at the thought of *any* risk of again producing such a malformed child, it is much kinder—and no less accurate—to present the case more optimistically by saying that 'there is a 95 per cent probability in favor of your having a normal baby next time.' Remember that modern young Americans are used to thinking in probability terms: every day they hear weather forecasters telling them of a '20 per cent probability of rain,' and when they watch baseball on television they regularly are told that the batsman is presently 'hitting .255' which means there is a 75 per cent chance that he will fail the next time he goes up to bat!

"When one gives a highly favorable prognosis, the parents' or prospective parents' relief and rejoicing is one of the most rewarding experiences in medical practice. Fortunately conditions with a good genetic prognosis—with recurrence risks of ten per cent or less—are more common than those with a bad genetic prognosis. But even in the latter situations, one should always try to find grounds for optimism. For instance, a child may be afflicted with neurofibromatosis associated with paraplegia: it is true that the risk of the recurrence of neurofibromatosis in subsequent siblings (or in the patient's children) is 50 per cent, but the mutant gene in this condition has a wide range of expressivity and affected family members with major nervous system involvement are much less common than those with just a few *cafe-au-lait* spots on their skins. . . .

". . . the family doctor usually proves to be a valuable ally to the genetic counsellor. He is likely to have had long and close contact with the family and a much more than superficial knowledge of their attitudes and emotional responses. Whenever possible, the genetic counsellor should enlist the family doctor's assistance in planning his approach to the consultants. It is in those cases where I have been able to closely collaborate with the family doctor that I have derived the greatest personal satisfaction out of the work of genetic counselling."

(We again wish to thank Genetic Counsellor Hymie Gor-

don, M.D. of the Mayo Clinic, Rochester, N.Y. for allowing us to use this foregoing material.)

Procedures, Costs and Other Factors in Genetic Counselling:

Unless you are fortunate enough to get your genetic counselling at a foundation or hospital where they are using research grants to secure material such as you and your family are able to supply, this is *not* an inexpensive procedure. It is now thought wise for a couple planning to be married to first undergo genetic counselling, especially when they know of a health problem in either or both of their families and-or ancestors; the cost is then doubled, of course, although the process may be either as simple as merely going through a series of questions and answers, or it may be as complex as the inclusion of a biogenetic and biochemical series of procedures for both. Of course, genetic counselling is automatically suggested when there has been a previous birth defect child in the family and you and your husband are concerned as to whether you should try for another child of your own.

Whatever your reasons for deciding upon genetic counselling, you may arrange to have it done in one of three ways: either as a referral from your family physician or obstetrician, as an outpatient at the nearest Birth Defects Center of the National Foundation-March of Dimes, or as a referral patient to a geneticist associated with a large medical hospital teaching complex, such as the one Dr. Hymie Gordon is associated with at Mayo Clinic in Rochester, Minnesota. However, be warned that this complex is seriously understaffed in genetics.

Should the condition about which you are inquiring be one of the genetic blood conditions such as sickle-cell anemia, Cooley's anemia, leukemia, hemophilia, Rh-blood factors, and so forth, then you might be directed to the Children's Blood Foundation, an independent, nonprofit organization functioning through the Division of Pediatric Hematology at the New York Hospital, Cornell Medical Center of New York City. If you are not able to visit New York, your own hematologist might contact the Foundation for the forms and instructions necessary to help them complete the research "long-distance" on you and your family, in order to come up with the biogenetic answers. Incidentally,

this Foundation happens to be one of those which does not charge for such counselling.

Dr. Denis Miller, this Foundation's Director, explains that diagnosing an inherited disorder requires a high level of sophistication by those involved, in order to understand the complicated biochemical nature of the genetic aspects of whatever genetic problem is being investigated. Besides all the family information and other data necessary, which were mentioned in the extracts by Dr. Hymie Gordon, Dr. Miller states that the steps required for assembling the counselling material entail the following:

(1) Collating the manifestations of the disease (providing you know or suspect what it is): clinical signs and symptoms, natural history of the disease, age at onset, sex incidence, complications of the disease, cause of death, special laboratory studies, x-rays, histology, cellular morphology.

(2) Establishing the precise mode of inheritance—construction of pedigree, identifying family members affected, determining if it is sex-linked, dominant or recessive.

(3) Discovering the general biochemical nature of the disorder—which entails identifying abnormal substances (metabolites) present within the blood cells or identifying other biochemical alterations.

(4) Identifying a defect in a specific enzyme or other protein gene product—the penultimate step.

(5) Determining the precise nature of the genic change, i.e., identifying the structural abnormality of enzyme function (decreased amounts of abnormal function).

Naturally, there are the other facts in the research inquiries. And there is always the big question for you and your husband to be prepared to answer: if you are told that it is definite that the child you may not even yet have conceived will have a difficult genetic problem—mental or physical—could you still take the chance of trying to have that child?

You must also know that thus far there are not enough genetic counsellors anywhere to fill the requests of all couples requiring genetic counselling. Even should you be fortunate enough to be put on such a waiting list, it may take weeks or even months before your own series of procedures is inaugu-

rated. After the information has been gathered, there will then be more weeks before the final genetic determinations are made—either by the counsellor and his aides, or by the counsellor and your own doctor.

Also, since some defects may crop up so rarely that they cannot be determined by genetic counselling procedures, you may feel you are wasting your time, to say nothing of the money involved. And yet, if you were to ask this writer, I would still say—do it! Find out everything you can about the possible baby you may have, before you have it, and even if there are no answers, you will know you have tried.

Of course, there is still another decision you may be called upon to make: Do you want to have your own child badly enough to conceive and wait a few months before using another of science's new procedures to learn whether or not the fetus is damaged—or can you put aside your natural desire to have your own child, and be happy just to have a child, one adopted but developed in another woman's womb, and can you still feel this is *your* baby?

Actually, you take the same chances many times with an adopted child that you do when you have your own. Many genetic defects, or even one-time birth defects, do not show up until months or years after the baby has been born. This can still be your child, it is still your problem to cope with, but your decision must be fast and firm before you either have a child or before you adopt one. Are you anxious enough for a child that you are willing to accept these averages, problems, or possibilities that are inherent in the creation of any and all babies? If so, you are very lucky—and that is why this book has been written for you.

Amniocentesis:

Amniocentesis is the seemingly simple process of injecting a special needle into the womb of the mother sometime between the 12th and 16th week of her pregnancy, and plunging it through to the protective wall between child and mother, to draw out amniotic fluid for examination and prediction of any possible genetic defects. What worries many obstetricians is that the incorrect insertion of the needle may accidentally hit the wrong place, despite x-rays in advance, causing irreparable damage to mother, child, or both.

Another thing to consider is that the later amniocentesis is

performed, the better the results for genetic prediction. However, it takes two more weeks for the cultures to be made from the fluid and possibly another week to study them and make up a report. If the report indicates a genetic defect in the fetus and the mother wishes a therapeutic abortion, she also has to remember that abortions are not usually recommended after the 12th to 14th week of pregnancy, and any performed later than that may also cause difficulties for the mother. Therefore, all of these questions must be resolved by the mother, father, and obstetrician.

The technique of amniocentesis was first developed in the 1930s, and the first large-scale studies were published in the March 12, 1970, issue of *The New England Journal of Medicine* by Henry L. Nadler, M.D., and Albert B. Gervie, M.D., both of Northwestern University Medical School and the Genetic Clinic, Children's Memorial Hospital. (Dr. Nadler is also Chairman of the Department of Pediatrics at Northwestern University.) These authors had performed 162 amniocentesis studies and felt the risk at 13 to 16 weeks was very low. But in an editorial in the same journal, Dr. John W. Littlefield stated this large study was the beginning of amniocentesis for prenatal genetic diagnosis to emerge as a legitimate scientific procedure. "Of course," he said, ". . . the ultimate decision . . . will be made by the families concerned, and will vary with their views, composition, aspirations, as well as consideration of local abortion laws and practices. The doctor should tell them that the likelihood of complications from amniocentesis seems very low, but that complications from the current methods for therapeutic abortion in the second trimester (after the third month) are not negligible. He must be able to inform them fully and accurately of the genetic risk that a child will be affected, and of the nature, available treatment and prognosis of the disorder. . . ."

Most of the articles in journals by genetics experts point out the wisdom of offering amniocentesis to all older pregnant women. Over half of those performed by Drs. Nadler and Gervie were done on women over 40, with this possibility of chromosome disorder in mind, since it is known that many of the births to such women result in Down's syndrome (mongoloidism).

In an interview that appeared in the September 28, 1970, issue of *The Journal of the American Medical Associ-*

ation, Dr. Nadler remarked: "All of the major chromosomal abnormalities are now detectable with this intrauterine technique. The procedure has proven valuable in determining the sex of the fetus when dealing with X-linked recessive orders such as hemophilia and muscular dystrophy. In cases of known carriers, a male child detected *in utero* would have a 50 per cent probability of expressing such a disease, whereas a female fetus would be either normal or only a carrier of the trait."

Roscoe O. Brady, M. D., assistant chief, Laboratory of Neurochemistry at the National Institute of Neurological Diseases and Stroke, Bethesda, Maryland, told physicians attending the Antenatal Diagnosis Conference in June, 1970, that more than 30 inborn errors of metabolism may be detected in the amniotic fluid cells or the fluid itself. All of the 9 known sphingolipid storage diseases, for example, can be diagnosed *in utero* through amniocentesis, and radio-labeled tracers or other biochemical testing techniques can reliably detect such problems as Tay-Sachs, Niemann-Pick, Gaucher, and Fabry's disease. However, relatively few investigators have had a great deal of practice combining microchemical, radioisotope, or other complex techniques with amniocentesis.

Scientists have now identified several hundred phenotypes or separately identifiable expressions of genetic disorders, according to an interview in the same September 28, 1970, *JAMA* with Victor A. McKusick, M.D., professor of medicine and director of the Division of Medical Genetics at the Johns Hopkins University School of Medicine. Dr. McKusick revealed that a new edition of his "Mendelian Inheritance in Man" was then being readied for publication and listed a total of more than 1700 distinct phenotypes—up from 1545 in the 1968 edition that had listed 793 autosomal-dominant, 629 autosomal-recessive, and 123 sex-linked phenotypes. Many of these conditions are rare, with only a few cases reported in medical literature. One estimate of the total frequency of congenital defects is at 5 per cent of all live births, and was made by Philip L. Townes, M.D., Ph.D., professor of anatomy (genetics) at the University of Rochester (New York) School of Medicine and Dentistry, at the 1969 genetic counselling symposium sponsored by the National Foundation-March of Dimes. Dr. Townes noted that many of the more common abnormalities such as cleft palate

and lip, clubfoot, and cardiac anomalies normally have recurrence probabilities in the neighborhood of 5 per cent or less. Such odds will probably not deter most families from having additional children. The chance of having children with certain hereditary problems can reach almost 100 per cent in some inbred families, which is why some states outlaw marriages between first cousins, and many geneticists try to discourage marriages even between second cousins, if the family history has too many birth defects in it.

Ordinarily, however, autosomal-dominant traits appear in half the male and female children of an affected individual. When both parents are carriers of an autosomal-recessive trait, there is a 25 per cent chance that their child will be affected. If sex-linked traits are involved, 50 per cent of the male children will inherit a disorder, and 50 per cent of the female progeny will be carriers.

Dr. Townes also stressed that more than 50 inherited disorders are now treatable if medical personnel identify the condition and begin therapy early.

"Although our screening programs for PKU (phenylketonuria) have revealed unsuspected heterogeneity," he said, "and thus unanticipated difficulties in diagnosis and evaluation of treatment, there is no doubt that early dietary treatment can be beneficial in PKU, galactosemia, congenital hypothyroidism, and many other inborn errors of metabolism."

Dr. Leon E. Rosenberg, associate professor of pediatrics and medicine at Yale University's School of Medicine, has warned physicians to be alert for the dozen or so hereditary metabolic defects which seem to be vitamin-dependent. Children with these disorders respond dramatically to specific vitamin therapy in doses ranging from 10 to 1,000 times their normal daily requirements.

Most geneticists are found in large, university-affiliated medical centers, many of which have grants set up by the National Foundation-March of Dimes, with associate grants set up by other large disease groups such as the National Cerebral Palsy Foundation, the ones for cystic fibrosis, and other crippling diseases, along with help in grants contributed by the federal government represented by the National Institutes of Health, Bethesda, Maryland.

If your doctor feels you need genetic counselling, there are three ways to find out if there is a counselling expert or center near you. Through an international guide listing these

services, which can be secured by your physician writing to the Professional Education Department, The National Foundation-March of Dimes, P. O. Box 2000, White Plains, New York 10602; through writing to The Public Relations Department, American Hospital Association, 840 North Lake Drive, Chicago, Illinois 60611; or through consulting the list of genetic counselling services in the United States (culled by the National Foundation from the International Guide) which follows this page. Many of these genetic counselling services include birth defects centers, and your doctor or pediatrician can find out whether they take care of the birth defect with which you are concerned. (See more on these birth defects centers in the section of this book that discussed the National Foundation-March of Dimes work.)

Now let us find out what is known about the most prominent birth defects that can develop.

THE NATIONAL FOUNDATION-MARCH OF DIMES,
P.O. Box 2000, White Plains, New York 10602
GENETIC COUNSELLING SERVICES
in the United States and Canada

The medical centers, universities, hospitals and clinics listed below provide genetic counselling services. Those marked with an "A" are able to provide amniocentesis, a new procedure, involving removal of fluid from the uterus, used to diagnose certain birth defects early in pregnancy. Directors of the Genetic Counselling Service may hold M.D., Ph.D., Sc.D., D.D.S., and other degrees, but for purposes of brevity we have listed them without citing degrees or specific titles.

Some of these facilities must be selective in the patients they assist because of lack of funds, time and manpower. Others operate under research grants to study a particular disorder. For this reason, most require recommendation from a practicing physician who is familiar with the patient's disorder and background. The cost of genetic counseling and of amniocentesis varies from area to area, depending upon facilities available and tests required.

This list has been compiled primarily from the International Directory of Genetic Services, third edition, compiled by Henry T. Lynch, M.D., Professor and Chairman, Department of Preventive Medicine and Public Health, Creighton

University School of Medicine, Omaha, Nebraska and published September, 1971 by the National Foundation-March of Dimes.

Copies of the entire International Directory of Genetic Services are available to physicians and others in the health professions on request from the Professional Education Department, The National Foundation-March of Dimes, P.O. Box 2000, White Plains, New York 10602.

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Facts About Various Specific Birth Defects

Parents who have lost a child, or have a living but seriously handicapped (either mentally or physically, or both) child, want more than reassurance. They frequently demand guarantees that no future child of theirs will be the same. Unfortunately, this guarantee cannot always be given. Science has not yet progressed far enough to learn everything about the cause and prevention of birth defects. National Institutes of Health scientists are working very hard, for instance, on a biochemical dissection of the human genetic disease called beta thalassemia, but primarily, it is still a fatal anemia that usually kills early in life, and the cause and cure are still waiting to be found in the distant future.

Dysautonomia:

This is a rare inherited disorder that renders its victims unable to feel pain, taste food, or produce tears when they cry. Dr. Alfred A. Smith of New York Medical College is now investigating the role of acetylcholine, a deficiency of which is thought to cause the disease. If successful, this research may eventually apply to other conditions such as mental retardation, myasthenia gravis, and other related neurological problems.

Phenylketonuria (PKU):

This is an example of the genetic inability of the body to use a certain kind of amino acid in the protein that is eaten. A build-up of this amino acid in the infant's blood will cause mental retardation. However, there has now been developed a diagnostic procedure through a blood test of the newborn, and treatment by restriction of the infant's diet can eliminate or, at the worst, minimize the mental retardation. Many

states require a PKU test of all new-born infants. The condition is caused when two persons marry, each having a PKU-carrying gene. Although about one in 90 persons carries such a gene, the chance of two of them marrying is about one in 8,000. Even then only about one in four of their children would show symptoms of PKU.

Mongolism (Down's Syndrome):

This is an inborn birth defect that is caused only in a small percentage of instances by a genetic mistake which can be inherited, such as an error in cell division, just before or at the time of conception. This puts an extra, separate chromosome into the fertilized ovum, making a total of 47 chromosomes instead of the normal 46. The result is Down's Syndrome. This kind of mistake is most likely to happen in a mother over the age of 40, who has never before had a child, or in one who has not had one in 10 or 20 years, probably because of the effects of aging on her reproductive cells, since they have been stored in her ovaries since the fifth month of her own prenatal life. Other women who may have been exposed to large amounts of X-ray may have a mongoloid child, or one may be born to women who previously have had such a child, while those whose blood serum contains thyroid autoantibodies are particularly prone to bear a mongoloid child. Male mongoloids are not known to have fathered children, and only a very few females with Down's Syndrome have ever had a baby, so this kind of chromosome mistake is almost never inherited. However, in some five per cent of mongoloid cases, studies show that the extra chromosome might have started with the mother. If this were so, there would be one chance in three that each of her children would be a carrier of this chromosome error but be otherwise normal, and one chance in three that each of her children would have normal chromosomes.

Maple Sugar Urine Disease:

Maple sugar urine disease is an inherited condition of abnormalities in body chemistry which cause mental retardation and death by the age of two years. There is a special blood test for this rare condition and the ailment derives its name from the maple sugar odor of the baby's urine. Science

is still trying to find a way to overcome its effects, as has been done with PKU.

Galactosemia:

This is a chemical birth defect causing mental retardation when babies drink milk. A blood test helps detect the disease in both victims and carriers.

Tay-Sachs Disease:

This is a so-called ethnic disease that consists of a defect in fat metabolism due to the absence of an enzyme, hexosaminidase A, in the brain. If an infant receives the TSD gene from only one of his parents, he will be healthy, but if he receives it from both parents, he will be a TSD baby, become inert, paralyzed, deaf and dumb, and die before his fourth or fifth birthday. Research has finally identified this missing enzyme which is so crucial to breaking down the fats. This offers some hope of controlling the problem, either by finding a way to supply the missing gene to the unborn infant's brain, or by replacing it after the baby is born. The TSD factor can now be found early in pregnancy through amniocentesis, giving the mother an option to terminate the pregnancy. TSD is known as a recessive genetic disorder.

Tay-Sachs disease is called one of the "ethnic" diseases because it has been traced back five centuries to the Ashkenazi Jewish people who lived around the area of the pre-World War I Lithuanian-Polish border, and still affects those of Eastern European origin or ancestry. Ninety per cent of the Jews in the United States are of that origin, and it has been estimated that one of every 30 persons carries this gene.

A simple screening test to discover carriers of TSD has been started in a two-year program, as of early 1971, by the John F. Kennedy Institute in affiliation with the Johns Hopkins University Medical School. Dr. Michael M. Kaback, pediatrician of Johns Hopkins, is director of this pilot program which may well serve eventually as a model for detection not only of TSD but of other inherited diseases, such as sickle cell anemia, cystic fibrosis, and others. The program will try to find carriers between the ages of 18 and 45, but if couples find out that they are diseased, the problem then is that many orthodox Jews will not have abortions performed.

This, of course, must be a personal decision of the affected couple.

Niemann-Pick Disease:

Although this disease does not show up in an infant until several months after birth, it is an enzymatic defect and an inherited disorder (discovered in 1966 by an NIH team headed by Dr. Roscoe O. Brady, Jr., a neurochemist) in which the afflicted babies are missing an enzyme sphingomyelianse, which breaks down something called "phospholipid sphingomyelin." However, the significant abnormalities caused by this defect make it important to have amniocentesis performed if any previous Neimann-Pick infant has appeared in the family.

There is another rare disorder of sphingolipid metabolism called Fabry's disease, that can also be detected through amniocentesis.

THE BLOOD DISORDERS:

Sickle Cell Anemia:

This is another ethnic disease, a product of the black man's African heritage. Originally prevalent across a wide stretch of Equatorial Africa, it has spread to those countries having large black populations and, with somewhat less frequency, to groups where blacks have mixed racially through marriage, i.e., Puerto Ricans of African ancestry and some Mediterranean peoples, such as Greeks and Italians. It is an inherited disease, serious and painful, sometimes fatal, a birth defect which affects about one in every 400 blacks in the United States.

Sickle cell anemia is caused by an abnormal recessive gene. About 10 per cent of the blacks in America have one abnormal gene for sickle cell anemia, and this is said to make them possess a "sickle cell trait," although they seldom have symptoms of the disease. When two possessors of the sickle cell trait do marry, however, there is one chance in four that each of the children born will inherit two of these abnormal genes and become victims of SCA.

In sickle cell anemia, the DNA in the pair of abnormal

genes carries incorrect instructions for the production of hemoglobin, a protein substance found in red blood cells. Each molecule of hemoglobin consists of two chains of 60 amino acids linked and entwined around each other in a precise order. The abnormal sickle cell genes cause the substitution of baline for glumatic acid in the beta chain of the hemoglobin molecule. Because of this infinitesimally small mistake, the chains of amino acids tend to lock together in an abnormal way when the normal amount of oxygen is not present. This causes the molecules of hemoglobin themselves to clump together abnormally and thereby distort the shape of the red blood cells so that they look more like tiny sickles than the usual indented discs. These sickled cells then tend to stick or mass together, often causing a temporary obstruction in the small blood vessels. This blockage cuts down on the amount of oxygen in that small area of the body producing more sickling of other red blood cells. A vicious cycle of more obstruction, less oxygen, more sickle cells, more obstruction occurs, leading to the damaging and even death of little local areas of tissue. The effects are bad—harsh joint pains, intractable leg ulcers, days of convulsions as a result of the affected central nervous system, sometimes damage to vital organs, and frequently unnecessary surgery for the kind of dire abdominal pains that suggest an inflamed appendix or a peptic ulcer, unless, of course, the doctor is the one treating the patient for his sickle cell condition, and knows the true diagnosis.

The periodic attacks of acute pain are called sickle cell crises, and can last for as long as four days, and general symptoms include weakness due to anemia, jaundice, malfunctioning of different organs, and leg ulcers. The disease often lowers resistance to infectious diseases, and apart from the antibiotics for the infections, the only treatment thus far for the sickle cell aspect is frequent transfusions, about once every month or so. Unfortunately, the transfused normal blood cells eventually become sickled, too, and the series of sickle cell crises start all over again. The hazards increase during pregnancy.

At the National Institutes of Health (which is the federal government's largest and principal medical research agency) a major research effort on sickle cell anemia is being carried on by three of the Institutes: the National Institute of Arthritis and Metabolic Diseases, the National Heart and Lung

Institute, and the National Institute of General Medical Sciences. There are a sprinkling of local sickle cell groups in various large cities and in New York the Children's Blood Foundation has programs for all the inherited blood diseases. Its work is discussed in another section of this book.

Individuals with only one abnormal gene for sickle cell anemia rarely have difficulties unless they are deprived of adequate oxygen during surgery, on plane flights, or while exercising at a high altitude, such as mountain climbing. The slight differences in the blood of those with a single sickle cell gene are enough to prevent malarial parasites from multiplying in the blood and thus can permit carriers of the sickle cell gene to be identified.

Experiments are currently being carried out on the use of hyperbaric chambers during periods of sickle cell crises. This has been found beneficial to adults, but not enough is known yet as to how helpful it can be for children.

Cooley's Anemia:

Another ethnic blood disease is Cooley's anemia (Mediterranean anemia) which basically affects children of Mediterranean descent, primarily Greek and Italian. It is a chronic, lifelong debilitating disease that affects normal body development, changes bone structure, enlarges the spleen to distend the abdomen, and limits a child's normal social activities. It causes social problems also, because skin discoloration and abnormal bone structure single out its victims. Unable to participate in physical sports, children with Cooley's anemia have unusually high absentee rates at school.

According to Dr. Denis R. Miller, Clinic Director of the Children's Blood Foundation, Cooley's anemia is caused by a genetic deficiency in the body which is unable to synthesize normal adult hemoglobin, the substance in red blood cells which transports oxygen through the body. It is not until the infant is about three months old that the first symptoms of Cooley's anemia show, for that is when the body usually starts to make adult hemoglobin. Instead, as the red blood cells break down the body cannot develop necessary new ones to take their place. Because the new cells given to the body with transfused blood also break down soon, the victim of Cooley's anemia must have another transfusion, then in a

short time another and another, continuing the pattern for the rest of his life.

The Children's Blood Foundation in New York City is one of the few with a transfusion clinic to give regular transfusions to its patients on an outpatient basis. Unfortunately, because of all these regular transfusions, children with Cooley's anemia develop an "iron overload" from the breakdown of both their own cells and the transfused cells. This iron gets into the heart, liver, and other organs, while the glands also become affected and become deficient in hormone production. There is now a medication, however, to help the body secrete this excess iron, and supportive hormone injections take over for the deficient glands, but many of the children still develop patchy discolorations on their skin because of the temporary excess iron condition.

The Children's Blood Foundation gives genetic counseling to parents to explain that if both parents have the genetic trait for Cooley's anemia, there is a 25 per cent chance their child may have the severe form of the disease. There is a 50 per cent chance the child will also be a carrier and his or her children will have it. Since 1961 a simple blood test given to people of Mediterranean descent has been able to determine whether they have the Cooley's anemia trait. Those with the mild form usually have no symptoms.

Hemophilia:

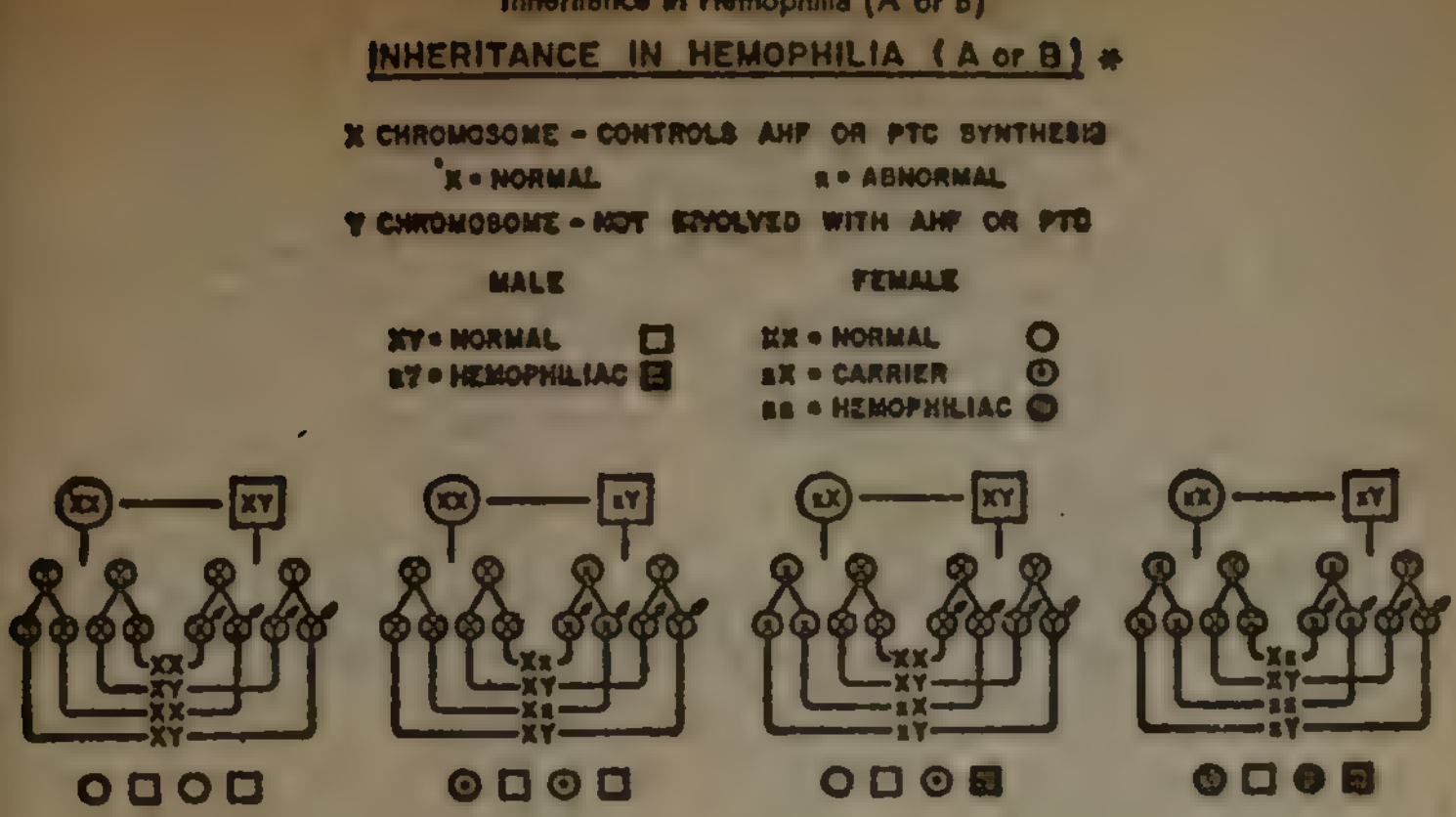
Hemophilia, the "Bleeding Disease," is a birth defect from a sex-recessive pattern. There are two forms of the disease: Hemophilia A (classical hemophilia, with a deficiency of AHF or Factor VIII) and Hemophilia B (Christmas disease, with a deficiency of PTC or Factor IX). The sex-linked inheritance pattern which is the same for Hemophilia and PT deficiency means that the genes which control the synthesis of AHF or PTC (also of color vision and many other characteristics) are located on the X chromosomes and when normal are indicated as large X, when abnormal as small x. Females have a pair of X chromosomes while males have one X and a Y. The Y chromosome is much smaller than the X chromosome and while it contributes little genetic information it is essential to the determination of the male sex. (See illustration which follows, through courtesy of The National Hemophilia Foundation.)

Approximately 80 per cent of hemophiliacs know of other bleeders in their families. In the 20 per cent without such positive histories, blood specialists (hematologists) assume that a mutation or change has occurred in the gene of the mother or of the ovum. Evidence also suggests such mutations are permanent and will be transmitted to future generations.

Hemophilia A, or classical hemophilia, is a hereditary disorder resulting from a lack or deficiency of antihemophilic factor (AHF). A complex protein, normally manufactured by the liver, spleen, and possibly other tissues, AHF is one of 11 known clotting factors involved in the intricate series of reactions leading to the formation of a blood clot. Hereditary deficiency states are known to exist for most of these factors and, with few exceptions, result in bleeding diseases of which hemophilia A is the most common. The defective gene responsible for AHF deficiency and hemophilia is carried by women who rarely suffer from the disease themselves. There is an approximately even chance that they will transmit the defect to their sons. The sons of a hemophilic man will be normal, but his daughters will be carriers and their sons will have an approximately even chance of inheriting their grandfather's disease. The coagulation defect of hemophilia can be corrected only by supplying AHF, and the correction is only temporary. Added AHF disappears rapidly from the blood; its normal half-life is only about ten to twelve hours.

According to the National Institutes of Health story sent us, until only a few years ago AHF could be supplied to the hemophilic patient only by transfusions of freshly drawn whole blood, or fresh or freshly frozen plasma. Except when needed to replace large amounts of blood or expand plasma volume, this involved transfusing a lot of unwanted substances along with the AHF and was a method that was particularly undesirable in surgery on a hemophilic patient, where blood volume might be expanded to dangerous levels.

In 1968 a major advance in the control of bleeding in victims of hemophilia was made by the American Red Cross under a research contract with the National Heart Institute, National Institutes of Health. A high potency, very pure concentrate of the AHF blood factor was prepared. The concentrate contains 40-100 times as much of the AHF antihemophilic factor as an equal volume of whole blood or


Inheritance In Hemophilia (A or B)
INHERITANCE IN HEMOPHILIA (A or B) *
X CHROMOSOME - CONTROLS AHF OR PTC SYNTHESIS
X = NORMAL
x = ABNORMAL
Y CHROMOSOME - NOT EVOLVED WITH AHF OR PTC
MALE
XY = NORMAL
xY = HEMOPHILIAC
FEMALE
XX = NORMAL
xX = CARRIER
xx = HEMOPHILIAC

* Courtesy of The National Hemophilia Foundation.

plasma. It is prepared from blood plasma through a relatively simple process developed by Dr. Alan Johnson of the American Red Cross Laboratory at New York University Medical Center, New York City. This technique enables the antihemophilic factor to be extracted from large batches of plasma. The remaining plasma can be processed to yield albumin, gamma globulin, fibrinogen, and other proteins useful for research and clinical medicine. The AHF concentrate is chemically stable and can be refrigerated for long periods without loss of potency. Its high purity minimizes the risk of side reactions and no hepatitis transmission was observed in test patients to whom it was given. It is manufactured for the Red Cross under contract, but not enough can yet be produced for mass, general distribution to all hemophiliacs.

About the same time, a similar concentrate, called "Method Four" antihemophilic factor (AHF concentrate) was developed commercially by Drs. K. M. Brinkous and Robert Wagner of the University of North Carolina in collaboration with the Hyland Division of Baxter Laboratories, Inc. Dr. Edward Shanbrom, then medical director of the Hyland Division, announced that this new concentrate would make it possible to treat about 90 per cent of hemophiliacs during visits to a doctor's office lasting only a few minutes. With transfusions of whole blood or plasma they

formerly had to be hospitalized. On a commercial scale, the Hyland product has 30 to 60 times the potency of plasma and can be injected intravenously through a syringe or drip bottle.

There are some 100,000 persons or more in the United States who suffer from this dread inability to have their blood clot normally and they know that any accident in which bleeding occurs may bring on severe crippling or even death.

In infants, the first sign of hemophilia may be delayed clotting after circumcision or bruising under the skin from minor bumps against the crib or other hard surfaces. Delayed clotting later during the first year might be due to a nick when cutting the nails, and puncture of the heel or earlobe for blood tests. Seldom does the eruption of baby teeth cause undue bleeding, even in a hemophilic infant.

Stubbed toes, skinned knees, cut fingers, and a bruised head are just part of growing up for most children, but to the hemophiliac and his parents these are the dreaded incidents that mean all kinds of anguish periodically. The childhood of most hemophiliacs is one episode after another of painful swollen muscles and joints that result from recurrent soft-tissue bleeding episodes and, in over 75 per cent of cases, represent the adult crippling stage of the disease. Repeated hospitalizations, poor school attendance, difficulty in applying ones self vocationally are just a few of its victims' problems.

Mothers of hemophilic children quickly learn that cold and pressure can work wonders with small wounds, whereas heat applied will only accentuate the hemorrhaging. A vial of the Hyland concentrate, containing 50 to 75 times the amount of AHF in a half-pint of plasma, costs approximately $40. Although the company recommends that its concentrate be refrigerated, it has been found that a hemophiliac may safely carry a small vial of it with him for a day or two in case a bleeding crisis occurs away from home.

Although the quantities made of their concentrate by both Hyland Laboratories and the licensee for the Red Cross are still not enough for all who require it, the hope of the National Hemophilia Foundation is that eventually there will be enough of it available so that this product can be used in much the same way that insulin is used for diabetics—on a

daily, regular basis for the lifetime protection and a passably normal life for all hemophiliacs.

Since the bleeding episodes in a hemophiliac can leave a great deal of pain in its wake, need for pain-killers is necessarily frequent. However, parents of hemophilic children should be cautioned about what they use for pain in their children. According to an article in the March 3, 1969, issue of *The Journal of the American Medical Association*, a clinical investigation by Dr. M. M. Kaneshiro and his associates at the University of Southern California School of Medicine in Los Angeles found conclusively that hemophiliacs of any age should not be allowed to use any medication containing aspirin. The variable effect of aspirin on bleeding time was found to be magnified in persons with bleeding disorders. Dr. Kaneshiro also announced that some people may respond to aspirin as normal patients do, with slight to moderate increases in bleeding time, but in others aspirin may profoundly prolong the bleeding time.

Therefore, rather than guess at whether your hemophilic child may safely ingest aspirin products, it is suggested that you keep these away from him, entirely. There are nonaspirin pain relievers, such as Valadol (Squibb) and Tylenol (McNeil) which are sold over the counter and without prescription, but before buying these, it is best to consult with your child's doctor, in case he has some other suggestion, instead.

Bilirubin:

Frequently in a premature infant, and sometimes in one which is full term, the skin becomes jaundiced during the first few days of life and an exchange transfusion of blood must be given to stem the jaundice. The yellowish hue stems from the excess bile pigments (bilirubin) in the blood. Full-term babies with jaundice due to a blood disturbance or Rh or O incompatability may also need a transfusion. Sunlight or artificial blue light has a decolorizing effect on some infants and may lessen the need for such exchange transfusions because light has the ability to speed the destruction of this chemical without producing toxic by-products. When premature infants are exposed to light therapy only the eyes are covered.

Too much bilirubin in the system damages the brain tissue. This occurs during the first week of life and an exchange transfusion is given when a blood test reveals high and increasing levels of bilirubin. Premature babies are born with a deficiency of an enzyme that normally eliminates the excess bile pigments from the blood.

Rh Blood Problems:

Still another problem that can affect the brain of an infant is erythromblastosis, in which the mother is actually "allergic" to her unborn child, which is one way of describing what happens when an infant develops Rh blood factor. Every year an estimated 250,000 women and their families risk having a baby affected by this problem. Thousands of such babies either die in the womb or are afflicted with mental retardation, congenital anemia, heart defects, jaundice, or spasticity. But conquest of this disease is now in sight.

Back in 1968 a serum was developed and manufactured by Ortho Diagnostics of Raritan, New Jersey and licensed as RhoGAM Rho (D) Immune Globulin (Human) to prevent Rh disease of the fetus and newborn, but somehow, medical authorities estimate, about one out of every four women needing the serum (or vaccine) is not receiving it, even though it can prevent Rh blood disease if it is given to a mother who is not already "allergic" within 72 hours of the abortion, birth, or miscarriage of each Rh positive baby.

What causes Rh blood disease? About seven out of every eight Americans have an element in their blood known as the Rh factor, and are called Rh positive. Those without this factor are called Rh negative. Whether a woman has this factor or not makes no difference to her own health, but it can make a difference to her children. If she is Rh negative and her husband is Rh positive, the child in her womb may inherit the father's blood factor, making the baby Rh positive, too. If some of this baby's blood gets into the maternal circulation as the child is born, the mother may react by forming antibodies to the Rh factor. The odds that this will happen rise with each pregnancy.

Every couple should have a doctor check their blood for the Rh factor, preferably at the time of taking the blood tests for their marriage license. If the couple proves to be that potentially dangerous combination of an Rh negative woman

and an Rh positive man, their family doctor (or later, the obstetrician) will take steps to protect any children they may have, provided the woman has not already become sensitized to Rh positive blood through a previous pregnancy or a blood transfusion. In such a case, the new serum will not be able to help her, as it is a natural preventive rather than a treatment of the disease.

New techniques exist by which physicians are able to learn, even before the baby is born, whether it is a victim of Rh blood disease. If necessary, the infant can be given a complete blood exchange transfusion while still in the womb, but usually an Rh baby is treated immediately after birth with exchange blood.

When used correctly, the Rh vaccine is definitely over 99 per cent effective, and, according to the manufacturer, will have no significant side effects on the Rh negative women to whom it is given. However, women with Rh negative blood must not conceal any previous pregnancies, miscarriages, or abortions from their obstetrician in telling him about themselves, otherwise, in not knowing the full story, he may unwittingly be prevented from helping both mother and child.

THE VIRUS INFECTIONS THAT CAUSE BIRTH DEFECTS:

Rubella:

An estimated 50,000 of our children died in the womb or were born with defects because their mothers contracted German measles (rubella) while pregnant during the 1964-65 epidemic. But, as a result of that epidemic, through participation in the Rubella Birth Defect Evaluation Project of New York University Medical Center, Louis Cooper, M.D., Professor of Pediatrics there, has worked with some 500 children and their parents and colleagues in the medical profession (as well as colleagues in a number of what he now knows to be allied professions, social service, special education and public health) to learn enough to be able to dispel the myths concerning rubella and to share what he considers are the major areas of ignorance concerning the rubella-damaged child. During our special interview with him, Dr. Cooper gave us permission to quote from a fulsome

article he wrote for *New Outlook for the Blind*, and one part is quoted here, while another part is quoted in another section on projects in education for the handicapped:

"Although rubella is one of the mildest illnesses caused by a virus, it is the only virus that regularly causes birth defects. This teratogenic potential was recognized first by an Australian ophthalmologist, Dr. Norman Gregg. In 1941 he noted an epidemic of congenital cataracts in his practice and by insightful history-taking was able to relate these events to maternal rubella in early pregnancy. Major strides in our knowledge concerning rubella followed the isolation of rubella virus and development of specific laboratory tests for rubella antibody. These laboratory breakthroughs occurred in 1962 in two laboratories, one at Harvard and the other at Walter Reed Army Medical Center. Because of these tests, we have been able to learn much about how rubella behaves and . . . what can be done to minimize or prevent future epidemics of rubella.

"Clinical rubella (the typical illness) begins approximately two weeks after exposure to someone who has the disease. Unfortunately, the person begins to shed virus in his nasal secretions as much as one week before he has any sign of illness. He may continue to shed virus for as long as two or more weeks. This is important because the person is frequently quite contagious well before he is aware that he is sick and he remains contagious for a prolonged period. Obviously, quarantine measures can not prevent spread of rubella because it is already too late by the time one knows whom to quarantine. Of importance to the pregnant woman and the developing fetus is the fact that the virus is present in the bloodstream for as long as a week before the onset of the rash.

"Laboratory tests are important because no doctor can make an unequivocal diagnosis of rubella based on appearance. Other viruses, which do not cause birth defects, can cause an illness which looks just like rubella. Even more difficult is the problem of 'subclinical rubella'—i.e., rubella without any rash or other signs or symptoms. This can only be detected by laboratory tests. Fortunately, in adults, rubella with rash appears to be more common than the 'subclinical' variety.

"The most generally useful laboratory test is called a rubella hemagglutination inhibition (HI) antibody test. If one

has never had rubella, then his blood serum does not contain this HI antibody. However, at the time the rash develops (or does not develop in the subclinical case) the HI antibody appears and persists for life. One bout of rubella infection appears to protect against subsequent reinfection. Of greater importance is the fact that a pregnant woman who has detectable rubella HI antibody in her blood at the time of exposure to rubella does not have to worry that her developing fetus may become infected.

"Unfortunately, we have learned that a history of past 'rubella' is totally unreliable. If we took antibody measurements from a group who thought they had rubella or whose mothers said they had rubella when they were children and a group of mothers who said they did not have rubella, we would find about the same incidence of antibody. This is caused by the factors mentioned previously, misdiagnosis and subclinical rubella.

"I believe you now can see how important it is that rubella antibody testing become widely available. In fact, one of our current goals is to see that this kind of antibody test be performed at the time a woman goes to get a marriage license or for her first prenatal visit. . . . If she is susceptible to rubella *and not pregnant*, she will be a candidate for (one of the) rubella virus vaccines. If she is immune to rubella (and 80 per cent of women of child bearing age are), then she can be reassured that she is not at risk. If she is already pregnant, and found to be rubella susceptible, she should be watched carefully, and then can be vaccinated immediately after delivery. Of course, the major thrust for rubella vaccine programs in this country (are) directed at young children (ages five to eight) who are responsible for epidemic spread of rubella. Elimination of epidemics will decrease the risk that pregnant women might be exposed.

"It is important that you know about these rubella antibody tests, because it is only through public demand that the tests will become widely available. . . .

"Rubella virus infection begins in the fetus during the first trimester or occasionally in the second trimester. The fetus remains infected throughout pregnancy and frequently for months after birth. The fetus can make antibodies of its own and if you measure antibodies in the blood of the newborn baby he has a good deal of rubella antibody. Much of the antibody was derived from his mother through the placenta,

but some of it is of his own making. If one can detect the presence of rubella antibody in a newborn infant that one can prove is of his own making (this antibody is rubella IgM which does not get through the placenta), then one is alerted immediately. Such a child must be followed carefully because the risk of rubella-associated defects is quite high.

"The fetus, once infected in the uterus, usually remains infected throughout pregnancy. In our experience, approximately 85 per cent of newborn infants with congenital rubella are still shedding rubella virus in pharyngeal secretions. By age six months, less than 50 per cent remain infected; and by age one year, the figure is less than 10 per cent. We are unaware of any child still infected after age two years. As long as the child is infected, he is contagious to those who have close contact with him. This is a special problem for hospital personnel, but is of little concern for the special educator who usually does not become involved until the child is at least a year old.

"Rubella and congenital rubella occur every year, but the effects of rubella are of greatest concern during severe epidemics, which occur unpredictably, but at approximately 7 to 10-year intervals . . . the major epidemic of 1964 . . . affected an estimated 30,000 infants. (It) was the first to occur after virus laboratory tests became available for definitive study of rubella and much has been learned from intensive studies of the 'rubella babies of 1964' which have been completed in many medical centers.

"Many women who had rubella in early pregnancy elected to have their pregnancies terminated by therapeutic abortion. Study of tissues from these abortions (by Dr. Marine and others) revealed that after virus is present in the mother's blood stream, the placenta becomes infected. From the placenta, virus travels through the umbilical cord to virtually every organ of the developing fetus. Since the infection becomes such a disseminated one, it is easy to understand how rubella can cause birth defects in many parts of the body. The virus does appear to have a predilection for certain target organs, however, and the eyes, brain, ears, and heart are especially susceptible to rubella infection and damage.

"The newborn baby with congenital rubella may appear to be perfectly normal or he may have numerous defects occuring singly or in any combination. Low birth weight (less than 5 and a half pounds) or birth weight one to two

pounds less than siblings is the rule. This is due to intrauterine growth retardation. Rubella virus slows down the rate of cell multiplication and therefore certain target organs have fewer cells than normal. Frequently, the rubella child, especially the more severely damaged one, will continue to grow more slowly than his normal siblings.

"Other transient neonatal manifestations include . . . bleeding into the skin because of a deficiency in blood platelets, hepatitis, anemia, and X-ray evidence of disturbed bone growth. These effects do clear in infants who survive.

"Other defects which are permanent include cataracts, glaucoma, heart disease, hearing defects, and brain injury. The cataracts may not be present or detectable at birth. It is not uncommon for a mother to be the first to recognize the presence of these lesions. Approximately half of rubella cataracts are bilateral. What are the implications . . . ? Obviously, if one leaves dense nuclear cataracts in place, a child is blind and has no useful vision. Furthermore, it has been learned that the rubella virus can lurk in these cataracts for years after birth, even long after the virus cannot be found anywhere else. (This is the exception to the two year rule mentioned earlier.) There are some ophthalmologists who believe the presence of the virus in these cataracts accounts for poor results in surgery and believe that one should delay surgery for the rubella-damaged child until he is three or four years old, until perhaps the virus is gone. I disagree. It is unfortunately all too frequently true that the child with bilateral cataracts is also deaf. If you leave the cataracts in place you have a deaf-blind child, a much more difficult problem than a deaf-low vision child.

"It would be nice if one could operate on the eyes of a child with bilateral cataracts the day after they are discovered. Unfortunately, this is technically extremely difficult and all too frequently results in the loss of the eye. In our experience, the compromise that seems to give the best results is to wait until the eyes get large enough for a surgeon to have a reasonable chance of a good surgical result. In practical terms, this means we are not operating on any cataracts until children are six months of age or older and most of our children end up having cataract surgery at about one year of age. . . .

"What kind of vision can one expect from an operated rubella eye? These eyes are frequently microphthalmic (quite

small) and not very good to begin with. Even if one could just dissolve the cataract without any further trauma, many of these children would have visual problems. With good surgery and proper fitting of glasses, a rubella child can have a good deal of useful vision, useful for ambulation and certainly enough for him to function in many low vision classes.

"(However) . . . it is extremely important that children with rubella cataracts be sent to eye surgeons who have broad experience with this disease. Experience with other forms of cataract is not a substitute for experience with the rubella cataract.

"In contrast to the rubella cataract where temporizing is appropriate, one must move quickly when congenital glaucoma is suspected. The diagnosis is usually an easy one—a big, bulging, cloudy cornea is hard to miss and increased intraocular pressure can be confirmed by tonometry under anesthesia. An infant with suspected congenital glaucoma should be referred without delay to an opthalmologist with broad experience with this disease. He will be able to distinguish this condition, which requires prompt surgery, from the transient corneal clouding occasionally seen in rubella syndrome. This latter condition requires no treatment.

"Unfortunately, the prognosis has not been good for . . . children with congenital glaucoma. . . . Delays and poor operative results have contributed to this problem. There is no adequate medication for this condition, and I would not hesitate, therefore, to send an infant hundreds of miles to insure that he would be in experienced hands.

"The most common ocular lesion in congenital rubella is a pigment abnormality in the retina which is usually referred to as 'rubella retinopathy.' This condition is easily detected by examination of the retina with an ophthalmoscope, which is a light with lens and mirrors which allows the doctor to look through the pupillary opening, directly onto the retina. The rubella virus causes clumping of the retinal pigments so that certain areas appear to be 'peppered' with dark granules and other areas are lighter than usual. . . . In most instances, this clumping does not interfere with vision. . . .

"The most subtle ocular defect in rubella is a most important one to detect. . . .

"Rubella heart disease almost always includes a condition called patent ductus arteriosus. The course of this . . . is quite variable. Many of our children have had no symptoms

from their heart disease and we have been able to defer corrective surgery until the third or fourth year of life, when it could be accomplished with relative ease and safety. Other children develop signs and symptoms of severe congestive heart failure during early infancy. Emergency surgery is frequently life saving, but congenital heart disease is the greatest contributor to the high mortality during the first six months of life. . . .

"Hearing deficits are the most common defects in congenital rubella. Among 376 children followed since 1964, 252 of them have significant hearing loss. Rubella deafness may be unilateral or bilateral, and has no characteristic audiometric configuration. The 252 children all have bilateral disease of sufficient magnitude to interfere with development of speech. Many of the children have visual and-or neurologic problems as well.

"In many ways the most difficult rubella deficits are those produced by the virus invasion of the developing brain. Many of our infants have had full-blown rubella encephalitis, and virus can be found in brain tissue at autopsy or in spinal fluid obtained during infancy. Since any area of the brain may be infected, the end result may be any degree or combination of intellectual and motor deficits. Some children have spastic diplegia such as is seen in typical cerebral palsy. Others have minimal or severe mental retardation. . . . Roughly three-quarters of our children clearly need some kind of special education. More than half would qualify as being multi-handicapped.

"What is the risk of congenital anomoly when rubella occurs during pregnancy . . . ? It is now clear that when rubella occurs during the first two months of pregnancy there is a very high risk of some kind of abnormality. During the third month the risk decreases, and falls off even more sharply during the fourth month. In our experience, rubella after the 18th week does not result in congenital defects. . . .

"It is important to realize that there are different kinds of defects depending on when rubella occurs. Cataracts, glaucoma, and heart disease are consequences of rubella during the first two months. Deafness and any kind of brain injury can follow rubella not only during the first eight weeks but any time during the rest of the first trimester and into the second trimester as well. This relates to the timing of development of organs. The heart and the eyes are well

formed by the end of eight weeks, but the central nervous system and the ear continue to develop throughout pregnancy. Since the virus infection persists throughout pregnancy and in early infancy, it should not be surprising that one can find damage to these organs when the rubella infection occurs fairly late. . . ."

The real danger of rubella is that the average person does not take it seriously enough. Many people know it only as three-day measles—an innocent-sounding alias, until you realize from Dr. Cooper's explanation that it has actually been happening for a week before the rash broke through. Almost everyone thinks of it as a harmless, childhood disease. But again, Dr. Cooper's explanation shows the harm it may cause to the pregnant woman who gets it.

The symptoms of rubella, ironically enough, are quite mild: low-grade fever, a rash starting on the face and spreading down the body, swelling of the lymph glands in the neck. These signs may be so slight that a pregnant woman may not even know she's had the disease—until her child is born with defects.

German measles (rubella) is not the same as "regular" measles (rubeola). They are two different diseases. The fact that a regular measles (rubeola) campaign for vaccination has also been conducted during the past few years has given many people the false impression that their children have already been inoculated against all types of measles. They have not. They must be vaccinated separately for German measles (rubella).

One further item about rubella vaccine. Women of childbirth age should have it only when the possibility of pregnancy in the following two months is practically nil. Dr. Cooper feels that each case must be considered individually. He also emphasizes that rubella vaccine is produced in cell culture and care should be exercised in administering it to persons with known hypersensitivity (allergy) to the species from which the series is derived (indicated in the labeling). For instance, he cautions that the vaccine contains a small amount of neomycin and should not be given to individuals known to be sensitive to this antibiotic.

During the personal interview we had with Dr. Cooper, he mentioned the handful of other infections known to cause birth defects, which include toxoplasmosis, cytomegalovirus, and syphilis.

Toxoplasmosis:

This is a fairly common protozoa that is very tiny. A large outbreak of it took place in 1969 at New York's Cornell Medical School in a number of students who had attended a lecture by Dr. Christian Barnard, the well-known South African heart transplanter. Afterward, the group went to the nearby snack shop for hamburgers and, because the meat was not well cooked, a short time later most of them came down with that virus.

Actually, by the time we reach adulthood, most of us have been exposed to the toxoplasmosis virus any number of times and might have had a case or two without knowing it. There may be no signs of a real infection, but it can cause a fever and you might complain of gastric intestinal problems without knowing it was toxoplasmosis. Only special laboratory tests can detect it.

Cytomegalovirus:

This virus is so nondescript that when it hits, Dr. Cooper advised, no one recognizes it for what it is. The effect seems to be bland on those having it, but if it happens at any time during a pregnancy (not just during the first three months as in rubella) it may cause severe brain damage, eye damage, and so forth, to the fetus.

The problem with cytomegalovirus is that it gives out so few obvious signs that the doctors still know little about it. They may find evidence of it when they are doing the immuno-suppression work to help keep an organ transplant (in a human) from being rejected. Such patients have their resistance to infection so lowered after receiving a transplant that even an attack of such a miniscule virus as cytomaegalovirus will exhibit more obvious signs than ordinarily.

Syphilis:

Few people seriously think of syphilis as a virus, yet it is an increasing problem among our liberated young, since it is a venereal disease that is widely out of control. Fortunately, there is a health law in New York and many other American cities that, during the first prenatal visit, the mother must be given a blood test for syphilis. If the test turns out to be ac-

tive, the pregnant woman is given penicillin, which will probably heal whatever might affect the unborn child, according to Dr. Cooper. However, if a pregnant woman with syphilis does not seek prenatal care until nearly the time of birth, nothing can be done about what then are definitely going to be the birth defect ravages upon the baby. These usually include both eye and ear damage, among other things.

ONE OF THE IMMUNO-SUPPRESSION BIRTH PROBLEMS:

Agammaglobulinemia:

A few years after the author's daughter (adopted) was born it was found she had a rare immunity defect—she had lost, or never had, many of the antibody factors in her blood with which to ward off infection. Fortunately, this was eventually cured by periodic injections of gamma globulin. However, another, much worse form of this lack of antibodies is called agammaglobulinemia, and is not so easily cured. Our daughter must have had at least some antibody factors, or the problem would have shown up within a few months after birth, when the mother's antibodies were no longer working for her.

We learned a great deal about the work of Dr. Robert A. Good, a blood disease expert at the University of Minnesota in Minneapolis, at the time of our daughter's problems, since it was he who referred us to a hematologist who found the remedy for her. Therefore, we were delighted to read of his later successes in bone marrow transplants detailed in the May 28, 1971, *Life* magazine story by Marion Steinmann. It told of a one-year-old boy from California who had already spent more than half his life in hospitals and had almost died three times. It was found that little Maurice had the rare genetic disease of agammaglobulinemia, and possessed virtually no immunological defense system. Because of this disease, the baby's body could produce no antibodies against invading bacteria and other infections and some of his white blood cells did not even work properly. In August, 1971, doctors at the University of California in Los Angeles decided to try the new procedure pioneered by Profes-

sor Robert A. Good (M.D.) of the University of Minnesota, which meant trying to give the baby an entire new immunity system by means of a transplant of bone marrow, a viscous red fluid containing primordial "stem cells" able to mature into the types of normal white blood cells he so desperately needed. The story said that until Maurice's transplant, similar bone marrow procedures on children with immune deficiencies like his had been tried only 16 times and of these only six had been successful. The first was done less than three years before by Dr. Good and Dr. Richard Gatti and the second was performed almost simultaneously at the University of Wisconsin.

"So far," said the *Life* story, "bone marrow transplants have worked only when the sick baby has been lucky enough to have a 'matched sib,' a brother or sister who has inherited his exact tissue type. Transplant specialists are now trying to extend the technique so that it can be used between individuals who are less closely related. . . . (These) have also been attempted, with mixed success, as treatment for a handful of other disorders (aplastic anemia, radiation sickness, Hodgkins' disease, leukemia) where the blood-forming mechanism either is congenitally defective or has been damaged by disease or drugs . . . as well as for such other blood disorders as thalassemia and sickle cell anemia. . . ."

One of little Maurice's sisters gave him some of her bone marrow in what was a fairly uncomplicated procedure. Thirteen-year-old Tami was given a general anesthesia in the hospital, after which various syringefuls of bone marrow cells were drawn from the front of her right hip bone. Little Maurice had only a local anesthetic. Then his sister's bone marrow cells were pumped through a catheter into his abdominal cavity.

"Because Maurice's immunity system was deficient, it could not reject Tami's bone marrow," the *Life* story explained. "But *her* cells were quite capable of launching an attack on Maurice and rejecting him, thus killing him. This type of reaction is called the graft-versus-host (GVH) reaction. . . . For 10 days nothing happened. Then, gradually, Maurice sickened . . . all signs of a severe GVH reaction. But then all by itself, the reaction gradually subsided, and he began to improve . . . the transplant was taking. Finally, two months after his transplant and nine months after he had first been admitted . . . Maurice . . . went home. . . ."

Diabetes:

In about one in four babies in the United States, there is the trait of diabetes. Clinical diabetes, or actual cases, is seen in about one in 2500 persons between ages one and 20; in one in 50 persons over 60 years of age.

Diabetes is a metabolic disorder characterized by the body's inability to handle sugar normally, leading to high glucose levels in the blood and the urine. The condition is related in some unknown way to abnormal utilization of insulin. Long-standing diabetics develop complications involving blood vessels, kidneys, heart, eyes and peripheral nerves. The disease comes in families but the mode of inheritance is not clear, although obesity does apparently predispose individuals to the disease.

Juvenile diabetes is frequently not apparent immediately at birth, but may show up in a growing child who is hyperactive, constantly hungry and thirsty, and gains weight easily. Pediatricians who suspect diabetes will give the child a glucose tolerance test of from four to six hours duration, to check whether his blood sugar remains high, low, or stable.

If there is a history of diabetes in your family, you should seek periodic check-ups before even thinking of pregnancy. If your child develops juvenile diabetes, even without a history of family diabetes, it can be handled positively with specific treatment. Generally, the child will have to use insulin injections daily for the rest of his life and be careful about his diet, but since there are known athletes and other important people in every field of endeavor who live to a ripe old age without suffering anything besides their diabetes, it is not a disease that should be treated as a doomsday problem. However, it should be treated, and it is generally agreed that a properly managed diabetic, even one who starts young, has a good prognosis. Without care, a diabetic will succumb sooner or later to various complications, including uncontrollable infection.

Pregnant Pre-Diabetics or Diabetics:

It has always surprised me how little importance lay periodicals writing about pregnancy ever give to the matter of the future mother who is a pre-diabetic or an actual diabetic.

They let it pass with such statements as "babies born to diabetic mothers are usually larger than other infants" or "mothers with diabetes or pre-diabetes have to be more cautious in pregnancy than those with other ailments," as though these statements are all such a pregnant woman has to know. They aren't. Actually, you should know what is facing you in either of these conditions, since a pre-diabetes condition can become actual diabetes at any time.

Before you visit your obstetrician, discuss the fact with your husband that the doctor may want you both to decide whether you may safely (for the baby as well as yourself) go through this or any other pregnancy. If your condition is dangerous for an extended pregnancy, you and your husband will have to decide whether the pregnancy should be terminated. It is that important.

According to a series of lectures on diabetes and pre-diabetes given in New York City in 1969 by Julius Pomeranze, M.D., internist and specialist in diabetes, diabetes mellitus is a chronic or lasting disease whose early stages are characteristically without complaints or symptoms of any kind, which is why frequently the sufferer does not realize he or she *does* have something wrong. During this noncomplaint stage, however, the presence of the disease is discovered only by chance during a routine physical examination or during the physician's search for the explanation of some seemingly unrelated sign or symptom, when he will have a specimen of blood and urine routinely checked and find high sugar levels in them. It is only much later that the obvious signs of increased urination, thirst, hunger, weight loss, and other symptoms of high blood sugar and disorder in the metabolism or handling by the body of carbohydrates, fats, and proteins do appear.

It is even true that the tendency or likelihood of a person to develop diabetes mellitus may be present long before any of the chemical signs such as sugar in the urine, elevated blood sugar or an abnormal sugar tolerance can be found. This is called pre-diabetes, and has been defined as the background of the person in whom sooner or later will develop the full-blown picture of the disease. The recognition of this stage of the disease is extremely important for both patient and doctor, because dietary regulation may not only prevent the chemical progression of the disease to its full-blown

form, but may also delay the degenerative changes in the eyes, blood vessels, kidneys, and other organs which are found in this condition.

As Dr. Pomeranze explains it: "Before you are born, you are pre-natal, yet already in existence; before you are diabetic, you are pre-diabetic—a state neither normal nor diseased, yet there. An underlying abnormality exists which may give vague hints of its presence and may show itself under certain specific conditions. An increased awareness of the condition pre-diabetes and the hints it gives of its hidden presence is the one available step forward in the possible prevention of the severity of the disease and its accompanying degenerative organ changes.

"The family history of diabetes is the first and most important characteristic of which the personal physician must be aware. An individual may inherit the weakness for this disease just as he may inherit the body configuration, the skin, the hair, or the eyes. Do any of your blood relations have diabetes? This search for a family history must include aunts, uncles, cousins, and grandparents as well as parents, for it may skip a generation. It is true, nevertheless, that a person may develop the disease and have no known affected relatives. Often this is because relatives have lost contact with one another, or do not reveal the presence of the disease. There is no way to predict when a person, liable to the disease, will develop diabetes.

"Among pre-diabetic females there is an increased tendency to frequent miscarriages. There is also an increased tendency in both pre-diabetic patients and in diabetic ones to give birth to babies larger than normal or babies with birth defects. It is, therefore, necessary for the married female to review with her obstetrician the birth history of her pregnancies. If any of these abnormalities have been present in a female with a family history of diabetes, the index of suspicion of pre-diabetes must be greater. It has also been demonstrated that pre-diabetic men, as well as diabetic men, father larger babies than do nondiabetics. This is not nearly as striking as in the females. The birth of a larger than usual baby should be a cause for looking for the presence of pre-diabetes in the mother and the father. . . ."

In the "Birth Defects Reprint Series" published by the National Foundation-March of Dimes, there was a dissertation on the "Infant of the Diabetic Mother" by Marvin Corn-

blath, M.D. and Robert Schwartz, M.D., the former a professor of pediatrics, the University of Illinois College of Medicine, Chicago, the latter associate professor of pediatrics, Western Reserve University, School of Medicine, Cleveland Metropolitan Hospital, Cleveland. The material was reprinted by the National Foundation from "Disorders of Carbohydrate Metabolism in Infancy," Volume III, Chapter 4, copyrighted 1966 and given special permission by the publisher, W. B. Saunders Company. We are using only a fraction of what the National Foundation used, just enough to give you an idea of what can result and already has resulted in births to diabetic mothers.

"If the mother was in the pre-diabetic phase, neonatal mortality was not abnormal, but total fetal wastage was increased significantly. . . . All authorities agree that if gestation goes beyond 28 weeks, survival is diminished for the infant from a diabetic pregnancy. The improvement in over-all survival rates in recent years—from 60 to over 80 per cent—appears to be the result of combined careful medical, obstetric, and pediatric management. . . .

"Factors related to pregnancy which may affect the outcome are toxemia and hydramnios. The incidence of toxemia is not greater . . . unless vascular disease is also present. . . . Hydramnios, an associated complication, may have an adverse effect upon the fetus. . . .

"Improvement in mortality rate has been attributed to a variety of factors of which intensive care by a medical team appears to be the most significant. . . .

"There is general agreement that insulin-dependent diabetics . . . benefit from interruption of pregnancy between 35 and 37 weeks' gestation provided the fetus is of reasonable size. Delivery prior to 35 weeks is associated with an excessive neonatal mortality. . . . Prior to the time of delivery, determined by obstetrician, internist, and pediatrician, the patient should be hospitalized and brought to a stable metabolic state. . . . All (obstetricians) agree that early delivery is much more important than the mode of delivery. . . . During induced labor and delivery, or during ceasarean section, intravenous administration of fluids with glucose and regular insulin may be necessary. . . . Maternal and umbilical cord vein and artery blood sugar values taken at delivery are useful baselines for future management of the infant. . . .

"The infant of the diabetic mother has survived diverse

metabolic alterations which have affected profoundly his intrauterine growth and development . . . but a significant number (40 to 60 per cent) have an uneventful neonatal period; the over-all survival rate is 80 to 90 per cent. . . .

"These infants often have, in addition to their obese . . . appearance, (problems) involving heart, liver, spleen, and hypertrophy of the umbilical cord. Not only are they overweight for gestational age, they have increased length as well. . . . It must be noted, however, that some investigators have suggested that there may be a more favorable outcome in the larger obese infants. . . .

"More recent data indicate that the excessive body weight is a result of increased body fat, rather than fluid. When starved and deprived of water during the initial days after delivery, they lose more weight over a longer period than do normal infants. . . . In the first 48 hours, urine volumes are larger than those from more mature infants of nondiabetic mothers. . . . Early feeding minimizes the weight loss and abolishes the difference when compared with nondiabetic controls. . . .

"One of the most serious neonatal complications is the respiratory distress syndrome. . . . The infant is irritable, with a 'complaining cry' and hoarseness. . . .

"The presence of respiratory difficulty in the initial hours after delivery may be related to intracranial anomalies or hemorrhage, cardiac failure, aspiration, pneumothorax, etc. In addition to careful physical examination, roentgen studies are necessary. . . .

"Both a respiratory and metabolic acidosis . . . are usually observed in the severe respiratory distress syndrone. . . ."

The paper went on to explain that many of these symptoms may come from a decrease in blood sugar after birth—hypoglycemia—and that with the respiratory distress may also be the hyaline membrane problem, for which today there is more treatment, and many cures, for the same illness which took the life of the late President Kennedy's newborn son, Patrick.

Bilirubinemia (about which we wrote in the part of this chapter on blood) also occurs more commonly in infants of diabetic mothers than in others, so it is obvious that even without physical deformities, there are enough problems associated with the possible live birth of a baby to a pre-

diabetic or diabetic mother to cause them to follow their doctors' rules and instructions for dietary and other treatment of their ailment before they plan their next pregnancy.

However, all of the experts we read agreed that babies born to diabetic mothers seem to have more congenital malformations than babies born to other mothers. And most of these malformations are taken up later in this chapter, since they do occur in other types of pregnancies and births also. It is wise to remember that having a large baby is never a sign that it will be healthy, because scientific investigation has proven that the smaller, seemingly slightly built child seems to have a built-in resistance to disease and other problems that the overweight newborn does not develop in many instances.

Cystic Fibrosis:

Although nobody has exactly pinpointed the cause of cystic fibrosis, researchers believe that babies born with this defect lack a certain key chemical, thus causing a fault in certain glands that make them act improperly. The mucus glands secrete an abnormally thick, gluey substance and the sweat glands produce an unusually salty sweat.

The most serious effects of cystic fibrosis result from the viscid mucus produced in the various organs. Normal mucus secreted by the lining membranes of organs is thin, slippery, and clear. In cystic fibrosis, the tenacious mucus creates two major problems for the affected child. One, it clogs the bronchial tubes in his lungs, interfering with breathing, and two, it plugs up his pancreatic ducts, preventing digestive enzymes from reaching his small intestine, with the result that the child is prey to bronchitis, pneumonia and other lung infections. The lungs, themselves, may suffer progressive, permanent damage, and, since digestion is impeded, severe malnutrition may occur if the condition goes untreated.

Cystic fibrosis is no longer considered a rare disease. The number of patients diagnosed as having this condition and treated for it has increased a great deal over the years. According to G. M. Harrison, M.D., associate professor of rehabilitation and pediatrics at Baylor College of Medicine, at a 1970 meeting of the Cystic Fibrosis Club in Atlantic City, N.J., he and his colleagues had found that measuring the amount of copper in nail clippings from newborn infants might

be able to provide a useful screening technique for cystic fibrosis. They did not know why the nails of cystic fibrosis patients contain more copper. But by using a neutron activation analysis technique, they found that the copper content of 118 cystic fibrosis patients, aged three months to 28 years, was 50 to 200 parts per million, but nail clippings from about 250 normal adult subjects contained only 2 to 40 parts per million of copper. The analysis of nail clippings from 850 neonates (newborns) showed the copper concentration was approximately the same as in normal adults—2 to 30 parts per million. During the period of investigation, one three-week-old infant suspected of cystic fibrosis was tested by both this new method and the old sweat electrolyte test. Both tests were positive.

Dr. Harrison and his associates consider the copper assay method is a lot simpler, less costly, and more suited to the mass screening of newborn infants than the presently-used test based on the sodium content of the subject's sweat. The infant's nails are trimmed with sterile surgical scissors and the resulting sample—usually 1 to 8 mg.—is irradiated with thermal neutrons. Results are usually available within 48 hours—often while the infant is still in the hospital nursery.

Cystic fibrosis is transmitted as a recessive trait, which means that both parents, even though healthy themselves, carry a gene for the disease. Children who receive a combination of two cystic fibrosis genes—one from the mother and one from the father—inherit the condition. Each time a pair of carrier parents have a child, there is a one-in-four possibility that the boy or girl will be born with cystic fibrosis. There are two chances in four that the child will have only one cystic fibrosis gene, and thus not have the disease but be a carrier. And there is one chance in four that the boy or girl will be entirely free of the gene and will have neither the gene nor be a carrier.

In the United States, cystic fibrosis is now one of the main causes of chronic illness in children and adolescents and one of the main causes of death in childhood, accounting for most of the chronic nontuberculosis pulmonary disease, for almost all pancreatic deficiency, and for at least some of the cases of hepatic cirrhosis. However, so much progress has been made with equipment and treatment of cystic fibrosis, that there are many more patients growing up from infancy to the young adult level, and the prognosis for even greater

progress and help with research and treatment of the disease is very positive. Formerly the disease was believed limited to the exocrine glands, but now researchers feel that all of the body cells may be involved in this generalized disorder. Furthermore, the different chemical changes in groups of cystic fibrosis patients suggest that the clinical picture of the disease might be due to two or more diseases masquerading as a single one. In March, 1971, outstanding investigators from various parts of the United States concluded a conference on cell and tissue culture in cystic fibrosis, under the auspices of the National Institutes of Health and sponsored by the National Institute of Arthritis and Metabolic Diseases, and the National Cystic Fibrosis Research Foundation. Dr. Paul A. di Sant' Agnese, chief of the National Institute of Arthritis and Metabolic Diseases' Pediatric Metabolism and conference chairman, said that the implications of this new research are far-reaching and give new insight into the basic molecular defect of cystic fibrosis. (This report appeared in the March 16, 1971, National Institutes of Health *Record*.)

Epilepsy:

No one really knows how many epileptics there are in the United States, because most of them still remain hidden, due to the fact that there are still many phobias and mistaken ideas about this ancient illness. Until the 18th century, epilepsy was generally seen as a demon possessing the mind and it was thought seizures resulted from the victim's efforts to free himself. For thousands of years the epileptic was little more than a human guinea pig used to test remedies which were absurd, disgusting, and often worse than the malady itself. Today, science (but not enough lay people) recognizes that epilepsy is not a disease but a term for the symptoms of certain neurological disorders which should be considered as separate entities for each individual. The common denominator is the interruption or loss of consciousness.

The cause of epilepsy can be in a baby at birth, such as brain damage or a tumor or lesion in the brain. The seizures are electrical brain-waves, shocks that may happen without relationship to any real cause or illness. There is no relationship between epileptic attacks and the intelligence or personality of the individual except where the seizures are due to brain damage. Some seizures will be a period of unconscious-

ness that is so brief that it passes quickly and will usually go unnoticed by others. Some seizures will consist only of a spell of staring or of slight muscle twitches. Others may be the more violent, convulsive seizures that require a handkerchief or stick under the tongue.

Statistics indicate that epileptic disorders are not hereditary. However, there can be psychological stresses produced by the way family members and other people (friends) react to the epileptic seizures, which may result in emotional problems for the patient also.

The public is confused about epilepsy primarily because it is one of the oldest, least understood ailments known to mankind, as well as one of the most maligned. People really do not understand the varying differences between those patients who have epileptic seizures as part of a general brain-damaged condition, usually present at birth, which leaves them severely handicapped, and those with plain epilepsy who are completely normal otherwise. There is even now medication that can keep the latter attacks minimal or else completely under control.

Many people believe most epileptics are mentally deficient or will eventually become that way. This is not so. Those with brain damage (the kind usually present at birth) are the only ones who are permanently handicapped, since nothing has yet been devised to make their brains whole again. But other forms of epilepsy can be helped, particularly the kind from operable brain tumors.

Epilepsy is an "irritative reaction" to injury—temporary or permanent—to the brain cells. There may be any one or more causes besides brain injuries and tumors, such as infections and-or body chemistry problems. The grand mal seizures are those with the big convulsions. The petit mal ones are slight and frequently not even noticeable. The minor motor kind are involuntary movements of a part of the body, such as a thumb or even the whole side, which may last for only a few seconds. Various other names are given to different kinds of epileptic attacks, but only a qualified neurologist can give an expert diagnosis of each type. Of the two million epileptics in the United States, it is estimated that one in 50 children has some form of it, and if not born that way, parents and teachers are asked by their PTA's to recognize the neurological signs that may require taking such children to a medical expert for diagnosis and treatment.

Children with epilepsy can and should be allowed to lead normal, productive lives, Samuel Livingston, M.D., the director of the Johns Hopkins Epilepsy Clinic, has stated.

"Of course," said Dr. Livingston in a story sent out by the AMA from a convention, "it's only natural for parents to be cautious in their care. . . . But the epileptic child needs no more rest than any other child, and to force him to lie down or take it easy indoors while everyone else is outdoors playing is not necessary, and is likely only to make the youngster feel put upon and unfairly deprived. . . ." He added that epileptic children can play with their friends, take part in many games and sports, and go to school. Many epileptic teen-agers and adults can hold down jobs, in most cases drive an automobile, marry and raise children.

If you now have or ever do have a baby with epilepsy, it is important for you to work with the groups trying to make a better life for epileptics. (See our section that discusses organizations and aids for birth defects problems.)

Among the problems, as the child gets older, are:

(1) A significant number of persons polled by the Epilepsy Foundation of America have proven they still have misconceptions about the disease. It has been found that over 10 per cent of them still object to their children associating with epileptics, 4 per cent still believe epilepsy is a form of insanity, and over 15 per cent do not believe epileptics should be employed.

(2) Persons with epilepsy are rejected by the U.S. military service and usually, but not always, are discharged should epilepsy develop while in service.

(3) Many children with epilepsy are barred by schools and universities, and there are too few special schools for children with epilepsy, except for the few who are retarded.

(4) It is estimated that up to 25 per cent of medically-controlled epileptics of normal intelligence are chronically unemployed and that employer resistance stems from (a) personal apprehension, (b) resistance of other employees, (c) discriminatory workmen's compensation laws, and (d) a general lack of proper training, counseling, and placement of employable epileptics.

(5) Automobile and health insurance are almost impossible for many epileptics to obtain and life insurance premiums are often much higher than the standard rate for nonepileptics.

If your child is fortunate enough to have remedial epi-

lepsy, then you and your family will have to be counted among those who band together to fight for his rights to education, employment, and a normal adult future.

Even if your child's epilepsy is the birth defect kind, you should work with the other epileptic groups to help all epileptics, in case some day there is a scientific breakthrough of even better drugs than there are now, to make your child one of those who can be rehabilitated for a better adult life.

Spina Bifida:

This is one of the tragic birth defects against which modern medicine is winning slow but hopeful progress. Literally, the Latin term means "cleft spine" and babies born with this condition have a spread-open split near the base of the spine as a result of incomplete formation and fusion of the spinal canal and failure of the protective bony vertebrae to close normally over the spinal cord.

Not long ago, most infants born with spina bifida died or became mentally or socially deficient. Today, the survival rate is up to about 60 per cent, largely as a result of better ways found, through research grants by the National Foundation-March of Dimes and the National Institutes of Health, of preventing hydrocephalus (enlargement of the head because of fluid accumulation), which often accompanies, or results from, spina bifida, or of eliminating it when it does happen, and methods of surgery for removing the fluid-filled cyst when closing the spinal opening.

Spina bifida, next after cerebral palsy, is the most common disabling disease of the nerves and muscles attacking the newborn. The incidence is variously reported as between 1.2 and 2.1 cases per 1,000 live births. Many of these children not only survive, but thanks to new methods of care and rehabilitation, are also able to go on to school and have hopes of eventually becoming self-supporting.

A soft, fluid-filled sac, or cyst, about the size of a small orange forms under the skin over the lower spine. The nerves and membranes of the spine, and sometimes a portion of the spinal cord itself, protrude and spread into the sac. Vital nerves thus are protected only by the skin and the walls of the cyst, rather than being sheathed in the tough bony structure of the spinal column.

The disarrangement of the nerves of the lower spine usually results in a loss of both movement and sensation in legs, and in severe cases, from the waist down. The excess accumulation of fluid around the brain (hydrocephalus) can result in head enlargement and eventually brain damage in spina bifida patients in whom shunt procedures may not work, and is related to the faulty pressures in a spinal canal which is abnormally open at its lower end.

If the soft sac over the lower spine is not removed shortly after birth and the opening closed, there is also the danger that this sac may rupture and expose the nerves to various types of meningitis infection. Babies which do die from spina bifida are the victims of hydrocephalus, meningitis, or urinary tract infections.

The combination of early surgery for the closing of the spine, antibiotics to ward off meningitis infection, and drain valves to remove the pressures of hydrocephalus fluid, has completely altered the outlook for survival of spina bifida babies. With modern aggressive treatment and special intensive care, between 31 and 33 per cent of such infants can eventually live normally. It is still too early to tell whether such babies will have a normal life expectancy, as well, since the first who received this combined form of treatment are now only in their 20's and 30's.

Obviously, treatment of spina bifida requires a great deal of teamwork between family doctor, pediatrician, neurosurgeon, a urologist to handle the chronic urinary tract infections which commonly develop and which may require surgery, and the orthopedic surgeon to treat the leg paralysis which is only rarely escaped with, perhaps, surgery or bracing and to reduce the hip dislocations which may occur. Constant nursing care is required to protect the skin and because of the lack of bladder and bowel control. With the aid of physical medicine specialists and physical therapists, many spina bifida children can be taught to walk as they grow older.

Experts believe that there is only a five per cent risk of a mother giving birth to a second child with a congenital malformation of the nervous system.

Only as recently as June, 1970, Ernest S. Mathews, M.D., announced at the AMA convention, that his department at New York University Medical Center had developed a tiny

strap-on high-pressure oxygen center averaging four inches in height, width and thickness, to drastically reduce spina bifida infection and lesion healing time.

In 1970, the Newington (Conn.) Children's Hospital and the Committee on the Care of the Handicapped Child, American Academy of Orthopaedic Surgeons, held a course on birth defects given by Dr. J. W. Sharrard of Sheffield, England. His main premise was that dislocation of the hip in myelomeningocele (spina bifida) is usually of paralytic origin rather than congenital dislocation, and transplantation of muscle will help overcome this condition which occurs in nearly half of all patients with spina bifida. In this way, he said, the hip can be maintained in its socket and a normal development of the hip joint should follow without bracing.

"If deformities have been corrected in early life," he said, "and a balance of muscle activity achieved, the correction will be maintained until growth is complete." Otherwise, he warned, failure to obtain balanced muscle activity would result in recurrent deformity.

He also told of an operation performed on the first day of life to help correct the curvature of the spine that happens to one out of 20 spina bifida victims. If this curvature is not corrected at birth, the deformity increases until the child is left in what can only be described as a jack-knife position. He cannot wear the braces that permit him to walk, he can't even lie on his back, and he will only be able to sit with his chest touching the front of his thighs.

Generally, most spina bifida medical experts recommend that early rehabilitation procedures are the only ones to prevent severe handicaps in later life. In various follow-ups of patients in the Myelomeningocele Clinic Program of New York Hospital, directed by Dr. Alfred L. Scherzer, it has been found that those patients who survived the complications of surgery and other problems as infants, have made satisfactory adjustments to their teen years. The program at New York Hospital includes special psychological sessions for parents and teen-agers, as well as the very important infant care.

Hydrocephalus:

Since this can occur without the spinal defect, but in cases of brain damage, it should be noted separately that the deaths

which occur happen primarily when the infant with hydrocephalus starts using shunts. These are taps inserted surgically into the head to try to draw fluid away from the brain. When successful they take the patient well into his lower school years or up to as high an age as eight or nine years, before they are removed permanently. The deaths are not due to either the shunts, themselves, or the hydrocephalus, but to those neurological or infectious situations which develop afterward.

Occasionally hydrocephalus will be arrested spontaneously in those patients in which it is only a temporary condition. Most of the hydrocephalus cases tend to become mentally normal or as nearly so as may be indicated by the success of the shunt procedures. What problems remain may include motor damage, necessitating a wheelchair and-or braces as the child matures. Indications are that when the shunt procedures are successful, the life expectancy of hydrocephalus patients is nearly normal.

The Cerebral Palsies:

Although this book is primarily on birth *defects*, some birth problems can be due to *injuries*, also or instead. Any of the cerebral palsies may be considered a general term applied to a group of permanently disabling symptoms resulting from damage to the developing brain that occurs before, during, or after birth, and that results in loss or impairment of control over voluntary muscles. Anyone may be affected by the condition regardless of race, economic standing, or environment, since anything which can damage brain tissue can cause cerebral palsy.

Unfortunately, about 25,000 babies are born with cerebral palsy each year, although some experts have raised that figure to possibly one newborn in every 150 live births.

The characteristics of cerebral palsy are:

(1) *Motor Involvement*—The major difference between cerebral palsy and other types of central nervous system disorders is the presence of motor involvement. These motor involvements include (when the child grows older) awkward or involuntary movements, poor balance, irregular gait, poorly articulated speech, tightness of muscles—all of which are caused by the brain's inappropriate regulation of muscle movements.

(2) *Associated Handicaps*—Although cerebral palsy denotes a person with a motor handicap, its relationship to the brain almost inevitably results in other types of handicaps as well:

a) *Mental Retardation*: from 50-75 per cent of children and adults with cerebral palsy are "mentally" retarded to some degree;

b) *Convulsions*: which may initially occur at any age;

c) *Speech Problems*: especially poor articulation (a result of faulty coordination of the speech musculature) affects 70 per cent of those with cerebral palsy;

d) *Visual Disorders*: including an eye turning in or out (strabismus) or visual acuity difficulty affecting 35 per cent of those with cerebral palsy;

e) *Hearing Impairments*: reported in 20 per cent of persons with cerebral palsy when the disorder is due to jaundice following birth;

f) *Shortening of an Extremity*: due to poor growth of a paralyzed arm or leg; if one leg is affected, the patient may compensate by tilting his hips—resulting in a curved back;

g) *Learning Disabilities*: conceptual and perceptual problems and language disorders may occur in children with generally average or above average intelligence;

h) *Behavior Disorders*: brain damage may result in less than normal adaptability to stress and anxiety; however, a major effect is poor parent-child relationships;

i) *Dental Abnormalities*: insufficient build-up of tooth enamel during prenatal development makes the person with cerebral palsy susceptible to dental caries and "irregular" positioning of teeth;

j) *Contractures*: joints such as the ankle, elbow, or wrist become permanently fixed in an abnormal or deformed position because of the excessive and unnatural pull of muscles around the joint.

There are a number of major types of cerebral palsy, each one just a bit different from the other. The five major types are:

(1) *Spasticity*—muscle spasms the most frequently occurring motor symptom of cerebral palsy;

(2) *Athetosis*—marked by constantly recurring, slow involuntary writhing movements of the arms and legs;

(3) *Ataxia*—walking like a drunken person;

(4) *Tremor*—similar to the fine tremulousness seen in adults who have Parkinsonism;

(5) *Rigidity*—muscles contract slowly and stiffly, leading to clumsiness.

The motor difficulty may involve all four extremities or it may be limited to just one, or an arm or leg on one side of the body may be disabled, which is the most common type. For one out of seven persons with cerebral palsy, the symptoms are so slight that except for some clumsiness they may be unnoticed. Others may be extensively handicapped, in a wheelchair and require assistance with the most basic activities of daily living, such as feeding and toileting.

While in over one-third of those affected with cerebral palsy the cause is unknown, some of the things which are known to cause this condition are:

(1) Prematurity.

(2) Anoxia (difficulty of the newborn in breathing).

(3) Complications of labor or delivery.

(4) Jaundice (yellowing of the skin) of the newborn due to uncorrected Rh incompatibility or incompatibilities of other blood types.

(5) Infections of the brain, such as meningitis or encephalitis, or an inflammation of the brain which may occur after routine cases of measles or German measles (rubella) and other viral diseases.

(6) Poisonings, such as lead.

(7) Accidents resulting in head trauma.

The general treatment goal for a baby born with cerebral palsy today is to help him function as well as he can within the limits of his basic handicap. While it may be impossible to alter the extent of his motor incoordination, it still is possible to train him to get the most out of what he has, by using treatment regimes through therapists which are geared specifically to improving the lack of coordination. It is still not known decisively, however, whether the improvement frequently seen after long-term therapy of this type is the result of treatment or simply the result of the child's growth and development.

Another major treatment objective is to prevent the secondary handicaps we have listed or, second best, to modify the harmful effects these conditions can cause.

Since damage to brain tissue cannot presently be repaired, there is as yet no cure for cerebral palsy as such. But the use of early and appropriate treatment and training does fre-

quently improve the mental and physical disabilities of cerebral palsy victims.

There has also been a great deal of research progress made so far. The virus causing German measles has been identified, as you know, and a vaccine licensed that undoubtedly will prevent many cases of multiple handicaps, cerebral palsy, deafness, mental retardation, and other handicaps resulting from fetal infection by rubella.

With major assistance from Public Health Service grants, a method of multiple exchange blood transfusions has been discovered for preventing kernicterus, a leading cause of infant death (approximately 1,000 deaths annually) and of cerebral palsy. Cerebral palsy from this cause has been reduced from 15 per cent to only one per cent.

The Rh-conflict type of cerebral palsy could be wiped out in the future through preventive use of the aforementioned Rh(D) Immune Globulin within 72 hours of delivery or miscarriage of an Rh positive baby by an Rh negative mother.

Physical and intellectual disability as a result of lack of oxygen at birth are being treated at birth chemically for anoxic newborns, with encouraging results. New drugs have also been developed which help ameliorate aberrant behavior (hyperactivity, impulsivity, anxiety) occasionally seen in children with cerebral palsy.

Last, family planning can reduce the incidence of cerebral palsies caused by pregnancies that occur when the mothers are too young, too old, or when their babies arrive too frequently.

(Note: Much of the above information on Cerebral Palsy came from a fact sheet compiled from the latest available information by the National Health Education Committee, Inc., 866 United Nations Plaza, New York, N.Y. 10017, which, in turn, used material from the United Cerebral Palsy Association, Inc., 66 E. 34th St., New York, N.Y. 10016; National Institute of Neurological Diseases and Stroke, National Institutes of Health, Bethesda, Md. 20014; National Easter Seal Society for Crippled Children and Adults, Inc., 2023 West Ogden Ave., Chicago, Ill. 60612, and figures from a survey compiled and published annually by "Drug Topics", Topics Publishing Co., Inc., New York, N.Y.)

It may be of interest to know that the Lesch-Nyhan syndrome birth defect has been found to be a tragic form of cerebral palsy that can be identified early in pregnancy, which demonstrates the feasibility of early prenatal diagnosis as a way of preventing this serious disease.

Lesch-Nyhan syndrome was first detected in 1964. Young male children who inherit the disease from their mothers have an uncontrolled behavior leading them to chew away their lips and fingers. They also have cerebral palsy, mental retardation, and ceaseless, uncontrollable movements of their hands. There is no effective treatment for these neurological aspects of the disease and the victims seldom live beyond puberty. Excessive uric acid in the urine of these children often leads to kidney failure. This condition which often contributes to their death can be prevented with the use of the drug allopurinol.

The Lesch-Nyhan syndrome results from an inherited biochemical defect, unlike most cerebral palsies which are less of a defect than an injury. It is caused by an absence of the enzyme hypoxanthine-guanine phosphoribosyltransferase (HGPRT) which causes the body to produce excessive quantities of uric acid.

The National Institutes of Health's National Institute of Arthritis and Metabolic Diseases supported in part a study which can lead to prevention of this disease. The investigators identified a male fetus with the Lesch-Nyhan syndrome at 21 weeks gestation, early enough for an abortion to be performed.

The researchers—at the University of California at San Diego, NIH, and George Washington University, Washington, D.C.—studied a woman who had previously had one normal child and one child with Lesch-Nyhan syndrome. When she was 18 weeks pregnant with her third child, the researchers withdrew a sample of amniotic fluid from the uterus (amniocentesis). A portion of the fluid sample studied revealed that the child was a male, which meant that he had a 50 per cent chance of having the disease. Another portion of the amniotic fluid was studied to determine the biochemical activity of the enzyme HGPRT. Cells from the fetus did not accumulate radioactively labelled hypoxanthine. This indicated a lack of HGPRT activity compared to normal human amniotic fluid cells. These normal cells with HGPRT activity did accumulate the radioactive compound. This bio-

chemical assay indicated the fetus had the Lesch-Nyhan syndrome.

On the basis of this information, the parents decided to terminate the pregnancy. The researchers then studied the aborted fetus, using two unaffected fetuses aborted for other reasons as controls. They looked for HGPRT enzyme activity in tissues from the skin, brain, genitals, and blood of the fetuses. They found no activity in the tissues of the fetus that had been identified by amniocentesis as having Lesch-Nyhan syndrome. By contrast, the two normal fetuses did have this enzyme activity, which confirmed the diagnosis that had been made during pregnancy.

The researchers suggest that children in cerebral palsy clinics and mental hospitals be screened for this Lesch-Nyhan syndrome. In this way, physicians can then identify female relatives who carry the gene for the disease and may pass it on to later children. With genetic counselling and amniocentesis in early pregnancy mothers who carry this gene can avoid having another child with the syndrome. Identification of children with the disease and their mothers will also enable physicians to examine other women in the family to see if they also carry the gene for the syndrome.

This study was conducted by Drs. J. A. Boyle, K. O. Raivio, K. H. Astrin, M. L. Graf, and J. E. Seegmiller at the University of California at San Diego; J. D. Scjulman at the National Institute of Arthritis and Metabolic Diseases in Bethesda, Maryland; and C. B. Jacobsen at the George Washington University in Washington, D.C. A report of this research also appeared in the August 14, 1970, issue of *Science* magazine, but the foregoing facts were sent in a press release by the National Institutes of Health, Bethesda, Maryland.

Hyaline Membrane Disease:

This is a disease present at birth, but primarily in premature babies and only recently has science found a cure for it. While the actual cause is not able to be pinned down patly, the most widely-accepted explanation given in a pathologic discussion in the April 15, 1971, issue of *New York State Journal of Medicine* is that it is caused by a decreased amount of surface active material (surfactant) in the lungs of affected infants. This decreased amount is blamed for atelec-

tasis, a prominent feature of the disease, which affects between 10 and 50 per cent of premature infants. Infants of mothers who have third-trimester hemorrhages or diabetes have a significantly higher incidence of hyaline membrane disease than the population at large. If normal respiration of the premature infant is observed for the first eight hours of life, hyaline membrane disease will not develop. Complications include bronchopneumonia as well as intracranial and other visceral hemorrhages. However, if the infant survives the first 72 hours, hyaline membrane disease usually clears without any further respiratory or other distress signs.

Recently a technique has been developed for the prenatal evaluation of the probability of a given premature infant being born with hyaline membrane disease, especially if the mother has a history of premature births in her children. This consists of performing amniocentesis and evaluating the phospholipid content of the amniotic fluid, enabling the doctor to predict when the requisite maturity of the lung tissue has been obtained. If birth occurs too soon, the obstetrician is at least prepared with the proper drug and machine procedures necessary for rescuscitation and eventual full recovery of the infant's breathing mechanism.

It was not until 1970 that the breakthrough came for this previously hopeless disease which caused the death of the late president Kennedy's last child, Patrick. "Hyaline" is derived from the Greek meaning "glassy" or "nearly transparent," and is a substance that seals off the air sacs in the prematurely-born infant. In 1970 two researchers—Dr. S. R. Snaklin of the University of Chicago and Dr. S. L. Wolfson of the University of Florida—recognized that more male infants died from this disease than female ones, and experimented with giving female sex hormones to such tiny patients. Not only was the entire number of such cases reduced, but the death rate for infant male patients was significantly lowered. It was found that the hormones helped best when injected any time within the first 20 minutes of life. Longer than that or, at the latest, longer than 40 minutes later than birth, the hormones did not help at all.

Another researcher believes he has found the answer to limiting this disease's fatal response. Dr. William F. Taylor of Los Angeles, suggests injecting an oxygen solution into the abdominal cavity as a temporary substitute for the inability of the lungs to deliver necessary oxygen to the infant in

the first 12 to 72 hours after birth. Unfortunately, more than a fourth of all premature infants have respiratory problems at birth, most of them involving the hyaline membrane.

Another breakthrough against the disease appeared in the March 6, 1970, issue of *Medical World News*, which told how the use of adult fresh blood in a baby with hyaline membrane disease who had hemorrhaged from the accidental dislodgement of a catheter in the umbilical vein, had resulted in the infant's recovery and eventually being able to go home perfectly normal. In another case, adult fresh blood was used in an exchange transfusion on a hyaline membrane baby, who had previously been thought hopeless, and improvement was marked after the exchanged transfusion.

Dr. Leonard D. Miller, professor of surgery at the University of Pennsylvania, later told the Society of University Surgeons meeting in Pittsburgh about these two cases he had attended in Philadelphia Children's Hospital, and stated that fresh adult red cells simply deliver more oxygen to tissues than fetal red cells which is why exchange transfusions of this type used with other standard therapeutic measures is believed to help sustain babies with hyaline membrane syndrome until it and other problems disappear spontaneously. Recovery usually occurs within three to ten days.

Marfan's Syndrome:

Marfan's syndrome (named for the French physician who identified this particular collection of symptoms as part of a single disorder), or arachnodactyly (literally "spider fingers"), is an inherited birth defect with such features as unusually long arms, legs, fingers and feet, loose joints, poor musculature, dislocation of the lens of the eye, absence of a fatty layer under the skin so that the person having the disease appears distressingly gaunt and underweight, and various heart abnormalities, including that of blood vessels, leading to such cause of death as an aneurysm (ballooning) of the aorta which ruptures without warning. The basic Marfan defect is thought to be a disorder of the connective tissue.

The pattern of inheritance in Marfan's syndrome is autosomal dominant, with variable expression, meaning it can occur in an individual who inherits only one abnormal gene for this defect from either father or mother.

Scientists suspect that Abraham Lincoln, known for his gaunt skeletal appearance, may have had Marfan's syndrome, but the symptoms may vary so considerably from individual to individual that in one person they are severe and-or multiple, and in another so mild that they can go undetected.

It is believed that about 15 per cent of Marfan cases are due to new genetic mutations. The odds for inheriting the disease or giving it to a son or daughter are one in two. Should a person not show any signs of having Marfan's syndrome himself, he may not be a carrier, even though a brother or sister may be.

Should there be any cases of Marfan's syndrome in your immediate family, it would be wise to have genetic counselling before deciding to have a child. If you are already pregnant, and unsure of whether the child is affected, your genetics counsellor will advise whether amniocentesis can give you the desired information early in your pregnancy.

Pompe's Disease:

According to a press release from the National Institutes of Health on February 1, 1970, scientists have been able to diagnose Pompe's disease, one of the rare inherited metabolic disorders, in an unborn child through a study of the amniotic fluid surrounding the fetus.

Pompe's disease, also known as type II glycogenosis or glycogen-storage disease, is a fatal inherited disorder of carbohydrate metabolism. Patients with the disease are deficient in an enzyme which is essential for the normal conversion of glycogen, the compound in which sugar is stored in the liver, into glucose, the basic sugar which nourishes living cells.

Most children born with Pompe's disease die from intractable heart failure before their first birthday. Until the advent of amniocentesis, there was no way to determine whether or not a child had the disorder until after birth.

Two of the Muscle Disorders:

According to Thomas Gucker, III, M.D., in the medical journal *Consultant*, April 1971 issue, muscular dystrophy actually includes some primary muscle diseases and some primary nerve disease and in many cases both in combination. "You are dealing with lesions of the final common pathway—

the anterior horn cell, the peripheral nerve, the myoneural junction and the muscle fibers. The muscles weaken and atrophy either (because) the nerve conduction function has been destroyed, or if the muscle fibers themselves have been damaged despite intact nerve conduction. . . .

"The most familiar variety (is) *Duchenne's pseudohypertropic progressive muscular dystrophy*. . . . Most likely the patient is a young boy with enlarged calves and waddling gait who falls frequently and literally has to 'climb up his legs' (Gowers' sign) to regain his balance. . . . In women carriers of Duchenne's dystrophy . . . the creatine phosphokinase level is usually elevated. . . .

"Still another variety is *true muscle atrophy of childhood.* In young children, this disorder produces Werdnig-Hoffman-type progressive spinal atrophy with muscle denervation and loss of reflexes, as well as hypotonia and, sometimes, muscle twitch. . . . In young children loss of muscle mass progresses rapidly to death within two years. In older children and adults loss of muscle mass progresses slowly with little reduction in life expectancy. . . ."

Mental Problems in Infants:

Retardation:

According to child health columnist W. G. Crook, M.D., early diagnosis of retardation in an infant is most important and may be able to eliminate a condition that could produce retardation. Parents should be made aware of what is expected in normal development, Dr. Crook advises, during the first six months of life. Some of the developmental "milestones" that the normal, full-term, healthy infant should pass during his first six months are:

6-8 Weeks—Baby should definitely notice parents and smile briefly when smiled at. He should also make slight "ah," "uh," and "eh" sounds.

8-10 Weeks—Follows faces or objects through an arc of 90 degrees, and smiles more readily.

12 Weeks—Coos, chuckles and follows objects from 150 to 180 degrees.

16 Weeks—Follows objects from one side to the other, across the midline, a full 180 degrees. He holds head steady with little bobbing. Very sociable, laughs out loud.

20 Weeks—Reaches for objects with both hands. Rolls over (back to stomach). Sits propped up for 15 to 20 minutes. Pats bottle.

24 Weeks—Rolls over (stomach to back). Turns his head toward sound.

"This is not to say that every baby who does not do each of these listed items at the stated age is retarded," Dr. Crook said. "However, such a baby should have a careful checkup to make sure everything is all right."

Autism:

One affliction of the brain is not necessarily considered damage so much as a psychosis. Autism will frequently develop in a baby's brain the first weeks after birth, regardless of how normal the parents are. It causes babies to live in a world of their own, not to seek affection or cuddling from their mothers and, by the age of two, to withdraw completely from outside influences. The smallest departure in routine can send infantile autistics into screaming rages. But they learn easily during their attentive periods, differing from brain-*damaged* children who cannot learn easily because of organic brain problems.

Autistic babies are *not* retarded. However, the January 26, 1970, issue of *Modern Medicine* listed 14 signs of autism in infants by Helen Clancy, Alan Dugdale, M.D., and John Rendleshort, M.D., of the University of Queensland, Brisbane, published in a 1969 issue of *Develop. Med. Child, Neurol.* An infant must show a minimum of seven of these symptoms before infantile autism can even be suspected and these must begin in the first three years of life. The 14 major manifestations are listed in decreasing order of frequency:

(1) Great difficulty in mixing and playing with other children.
(2) Acting as if deaf, with no reaction to speech or noise.
(3) Strong resistance to any learning, either new behavior or new skills.
(4) Lack of fear about realistic dangers, such as fire or dangerous height.
(5) Resistance to even the smallest change in routine, which may result in excessive anxiety.

(6) Preference to indicate need by gestures, although speech may or may not be present.
(7) Laughing or giggling for no apparent reason.
(8) Not cuddlesome as a baby—either holds himself still or clings limply.
(9) Marked physical overactivity, such as waking and playing for hours during the night, but remaining full of energy the following day.
(10) Persistent tendency to avoid eye contact by looking past or turning away from people, especially when spoken to.
(11) Unusual attachment to a particular object, with no regard for its real use.
(12) Spinning of objects, especially round ones, and distress if interrupted.
(13) Repetitive and sustained odd play, such as rattling stones in a tin.
(14) Standoffish manner and little communication with other people.

The Australian investigators emphasized that these manifestations provide only a presumptive diagnosis of autism and should be supported by a full clinical history and examination, careful assessment of family interpersonal relationships, electroencephalogram, urine examination for amino acids, and psychological testing using all relevant tests.

According to the April 6, 1970, issue of the *Journal of the American Medical Association*, allergies may also lead to minimal brain dysfunction in children. A pilot study with 20 children by Fred J. Kittler, M.D., supports this contention, and Dr. Kittler told a group at the American College of Allergists in Miami Beach that a larger study is now under way. He cited characteristics of a type of "tension-fatigue syndrome" which suggests food allergy as a cause. Such a patient is "an irritable, sluggish and disinterested child who has unpredictable behavior," said Dr. Kittler, clinical assistant professor of pediatrics at the University of Arkansas Medical School in Little Rock. "He is tired after a full night's sleep. . . . The classic hay fever, asthma, and eczema symptoms may be absent, but allergic findings (of) periorbital puffiness and dark circles under the eyes, generalized pallor and (runny nose) are usually noted."

In his pilot study, Dr. Kittler noted that diet restrictions

not only improved behavior and learning performance, but even led to correction of abnormal electroencephalograms.

"Minimal brain dysfunction" or "learning disabilities" are words used to cover a wide assortment of problems with many differing causes. Some of these are known to be birth defects, because they result from prenatal injuries or infections or genetic differences already found present in such children when they are born.

At least one kind of reading disability is thought to be genetic in origin, probably with a sex-linked recessive pattern of inheritance, because it affects males more often than females. Premature birth also is frequently thought to cause learning disabilities, especially when the prematurity is not the time in which the baby develops in the womb, but in its low birth weight.

Other forms of minimal brain dysfunction may simply be quite mild variations of cerebral palsy or epilepsy or an unrecognized inborn error of metabolism. Many youngsters with learning disabilities are known to have a medical history suggesting such prenatal damage as that caused by rubella during the mother's pregnancy, but they were fortunate enough to have escaped the other, more serious mental and physical results from the rubella infection.

Researchers believe that another cause of brain damage in the newborn may well be convulsions, since brain-wave tracings in such babies may predict their chances for normal development. These EEG readings are a more accurate predictor than either the cause or the clinical symptoms of the convulsions which, during the first few weeks of life occur in many infants and may foreshadow serious neurological disturbance. Other studies have attempted to predict a child's future development on the basis of cause, clinical symptoms and EEG readings during a convulsion.

In a study of 137 newborns with convulsions, released on September 15, 1970, by the National Institutes of Health, Arthur L. Rose and Dr. Cesare T. Lombroso found, at The Children's Hospital Medical Center in Boston and at Harvard Medical School, that those with a normal EEG have an 86 per cent chance of normal development at age four years. Those with mild EEG abnormalities have a 69 per cent chance of normal development and those with gross abnormalities, a seven per cent chance. This study was supported in part by NIH's National Institute of Child Health

and Human Development. During the study, the researchers observed all the full-term normal-weight infants in the hospital nursery, enabling them to learn about each baby's condition preceding the seizure and to carefully note the baby's clinical symptoms during it.

They found that after four years, 50 per cent of the 137 newborns who had had seizures were normal, healthy children, but 30 per cent had some neurological complications (mostly mental retardation) and 20 per cent had died, some from causes unrelated to the seizures. They noted that neurological signs in an infant during a convulsion were not an accurate predictor of future development. Some infants with serious neurological symptoms recovered completely, while some with milder signs had lasting neurological problems.

The primary causes of convulsions were hypocalcemia (low calcium levels), injury to the head at birth, infection, brain malformations, lack of oxygen during or before birth, longer than normal gestation, pre-eclamptic toxemia (a condition in late pregnancy characterized by high blood pressure and edema (swelling) and hypoglycemia (low blood sugar). They recommended that physicians use the technique of intravenous infusions to tell whether a metabolic imbalance is causing the seizure—even before an anticonvulsant drug is given. Since seizures are not usually violent in newborns, the researchers believed it most important to discover the cause. Excessive administration of drugs at the seizure's onset may not be necessary and may also mask the underlying cause of the attack.

During March, 1971, writer Joan Beck, who does family-oriented columns for the *Chicago Tribune*, attended the International conference of the Association for Children with Learning Disabilities, and wrote up its findings for the March 14, 1971, issue of her paper, parts of which we give you here:

"Some seven per cent of children seem to have learning problems and behavior problems caused by neurological differences, most of them coming from poverty areas since children born into poor homes are more likely to be exposed to possible causes of learning disabilities—such as prematurity, infections, or malnutrition during prenatal life and complications of pregnancy and childbirth.

"Boys outnumber girls with learning disabilities and teachers during the past decade have found more than 320

different symptoms of these disabilities. Some children may have only a few of these symptoms, but most of them at least have some. The primary symptoms are:

"*Perceptual Disabilities*: Although his eyes and ears check out as normal, the child may not see or hear what normal individuals do because of a difference or dysfunction in the brain or nerve pathways to the brain. Or his tactile and kinesthetic senses may not operate adequately. Or his brain may not be able to relate this sensory input normally. He may have difficulty judging distances, separating background from foreground, or knowing exactly where all parts of his body are in space.

"*Poor Coordination*: A child with learning disabilities is often clumsy, awkward. He can't make his hands and eyes work together well. He can't draw a man or print letters or ride a bike or do as well in sports as classmates. Often, he isn't definitely right- or left-handed. He may confuse right and left.

"*Abnormal Activity*: Typically, a youngster with learning disabilities is hyperactive (or, to give it the scientific name, hyperkinetic). He can't sit still, even when he tries. He talks in torrents. He jumps from one activity to another. He can't seem to stop making purposeless motions. He sleeps less and more restlessly than other youngsters his age. When his activity is measured scientifically, it can be four or five times more than average for his age. Or, in some cases, a youngster with learning problems may be abnormally slow and sluggish in everything he does.

"*Short Attention Span*: Many youngsters with learning disabilities can't concentrate. They can't screen out unimportant sounds or sensory stimuli to focus on one activity. Such a child may not be able to fix his attention on the words he's supposed to read because he's distracted by the other words on the page, by the illustration, by the other children in the room or the car outside the window.

"*Impulsiveness*: Not only does a child with learning disabilities tend to notice stimuli all around him, he often has to react immediately and without thinking to those distractions. He tends to be reckless, explosive, unpredictable, uninhibited.

"*Immature Development*: Many children with learning disabilities look and act young for their age and their intelligence. They may be late in toilet training or lag behind in learning to walk or talk or show readiness to learn to read.

"*Emotional Instability*: It's typical of a child with learning disabilities to have tantrums over trifles. He may panic easily, be quick tempered, difficult to live with. He may get too excited to play with more than one youngster at a time.

"*Language Disorders*: Speech development is sometimes slow in children with learning disabilities. Some stutter. Others may mispronounce several sounds long after the age of five or six. Some children have trouble understanding verbal directions.

"*Specific Learning Disabilities*: Typically, these youngsters do markedly better in some school subjects than others. Over-all test scores may be normal or higher, but sub-test scores vary greatly. Reading, writing, math, and spelling tend to be difficult for these youngsters, but many do well in some areas and fail in others.

"*Neurological Signs*: Irregularities on the electroencephalogram can be found in more than half of the children with learning disabilities—if the EEG is read by a skilled neurologist. But almost all of the youngsters have what doctors call 'soft' neurological signs. Their reflexes may not be quite normal. They may have trouble focusing their eyes on an object and following it as it is moved. Physicians sometimes disagree on how significant these minor findings really are."

Who should diagnose learning disabilities in a child? Joan Beck's article ends on this note: "Too often in the past, it's been a parent who recognized a description of his youngster's problems and had to insist to a teacher or a school psychologist or a physician that such a diagnosis be made. Or it may be a teacher who first calls attention to the symptoms of learning disabilities because she is in a position to compare the child's behavior with others his age."

Congenital Birth Defects (Physical):

Among the most useful techniques for determining an infant's ability to survive is the Apgar Score, based upon five objective signs—heart rate, respiratory effort, muscle tone, reflexes and color—all judged within one minute after birth. A low Apgar Score and extremely low (or extremely high) birth weight are danger signals that cannot be ignored and call for prompt treatment.

Sometimes there are observable clues to internal physical defects. The normal umbilical cord, for example, has three

easily seen blood vessels. When there are only two, further investigation may reveal a defect in the kidneys, heart, or other internal organs. Other methods are used soon after birth to identify hidden structural defects such as the lack of a connection between the esophagus and the stomach, one of many conditions that can be corrected by surgery.

Some abnormalities are so slight that they are but minor inconveniences. In color blindness, for instance, it means only that the individual cannot tell red from green. Or a child born with an extra finger or toe suffers no great problem, since it can be removed surgically.

On the other hand, there are a great many other congenital malformations (physical birth defects), as well, including:

Birthmarks: While many of these are unimportant, the disfiguring ones are reddish or wine-colored patches, more purplish than pink, consisting of numerous small dilated blood vessels and sometimes referred to as "port wine marks." True blood vessel tumors are rarer than these. They are elevated and may occur on any part of the body. Their cause is unknown and skilled plastic surgeons are able to remove a great many of them. Skin grafts are frequently used, and a relatively new technique for large marks is to tattoo normal skin colors right over the wine-colored area.

Cleft Lip (*harelip*) *and Cleft Palate*: About one in every 1,000 babies in the United States is born with a cleft lip. Two-thirds of these also have cleft palate. The frequency appears higher in white than in black populations. When the embryo is about six weeks old, swellings that will become the upper lip have not yet met. If they do not fuse at the proper time, the gap will remain and the baby will have a cleft lip.

About one in every 2,500 babies has a cleft palate without cleft lip, so that it appears the two conditions are not genetically related. A cleft palate may simply be a hole in the roof of the mouth, but sometimes that hole will also include the nasal area and part of the nose is not formed as well. More boys than girls have harelip and more girls have cleft palate.

These conditions are sometimes related to genetic defects. In experiments with animals, it appears that the environment in the uterus, such as the position of the embryo or the blood supply, may be a factor, and some drugs given during pregnancy are also under suspicion. There are now many new operative techniques for repairing the defects. Harelip can be

repaired in the first few weeks after birth and cleft palate before the child is 14 months old in most cases. There are also special nipples used for bottles in cases of cleft palate, so the baby can drink liquids almost normally.

Clubfoot: About one in every 250 babies in the United States is born with a clubfoot, a condition in which the foot turns inward or outward and is fixed in a tiptoe position. It possibly occurs due to the position of the child in the uterus, although malformation of the limb bud may also be the cause. Mild deformities respond well to shoe splints worn at night. Simple braces and corrective shoes may be needed. More serious clubfoot will often be associated with other defects, such as neurological ones of the cerebral area and the spine. Treatment must begin early and is prolonged, because the condition tends to recur. The muscles and ligaments must be stretched and the bones realigned. Plaster casts can often correct the condition, but sometimes surgery is required.

Congenital Heart Defects: About one in every 200 babies in the United States has a congenital heart defect, of which some are so slight as to cause little strain on the heart; others are fatal. In some of the abnormalities, the baby appears blue. Few causes are known, but one is certain—German measles (rubella) in pregnancy. An ever-increasing number of these congenital heart conditions are repairable by surgery.

Congenital Dislocation of the Hip: This is a rather common defect, frequently taking place at birth, especially if it has been a difficult or breech birth, and the infant's legs have been yanked at or pulled to effect delivery.

There is a whole range of abnormalities which can occur in the hip joint, since not always will the ball-shaped head of the thigh bone fit correctly into the capsule or socket of the hip bone. Sometimes, the head of the thigh bone just slips too easily in and out of the socket, which is a simple dislocation, but if it happens because the parts of the joints are not shaped correctly and will not fit easily or work smoothly, then there's real trouble. If the condition is not caught during the postnatal stay at the hospital, it is usually noticed by the pediatrician when the baby tries to walk and has a characteristic limp of hip dislocation. If it is caught early—during the first three months of the baby's life—there is little difficulty in correcting it with splints or a cast to hold the hip in the correct position. However, it must be remembered that the medical rule of thumb concerning correction of congeni-

tal hip dislocation is that for every month of delay after the third month of life, the baby will have to be kept one more month in corrective splints, or the procedure may even involve putting the little one in traction for a while and then having to use a plaster cast on the hip for several months longer. This is because any delay to correct the dislocation will cause the head of the thigh bone to move further out of place, especially as the growing child's weight becomes heavier on the involved hip and leg. Muscles also become shorter, the ligaments stretch and become deformed and generally speaking, the tissues involved in the affected hip joint are not able to develop properly, which accounts for eventual crippling. Medical experts believe, however, that there can be good correction of congenital hip dislocation if proper treatment is started before the age of three. After that, major surgery most likely is needed, and may not turn out 100 per cent favorably, as there will be some amount of foreshortening of the limb, a great deal of limping and transitory flashes of pain whenever the child strains to walk normally but cannot.

Congenital Urinary Tract Defects: About one in every 250 babies in the United States has one of the many different types of congenital urinary tract defects involving the kidneys, ureters, bladder, and-or genitalia. Organs may be absent, fused, or obstructed. Some causes are known, such as certain hormones given during pregnancy, and there is also some hereditary tendency toward such conditions. Most of these problems can be corrected by surgery.

Missing Limbs: While this is generally considered very rare, congenital amputees are born with one to four limbs missing or seriously deformed. While the cause is said to be unknown, it is recognized, since the thalidomide tragedy of a few years ago, that when pregnant women are given certain drugs, they can produce anatomically-deformed children. Great strides have since been made in prosthetic or artificial devices to enable such children to lead nearly normal lives. The emotional problems of the affected parents and children add up, of course, but are being overcome to a large extent through mental health programs.

Today, obstetricians are very careful about the medications pregnant women take, except where it is a matter of saving the life of the mother during a crucial illness or infection for which medication must be used.

Osteogenesis Imperfecta (*Brittle Bone Disease*): This is a rare, inherited disease that until recently was thought to have resisted treatment. One of the signs of the disease is sweating, another is that the child's bones are so brittle that a heavy touch, a slight fall, or even attempting to stand or walk, can cause fractures throughout the bony frame of the child.

At a 1971 meeting of the American Academy of Orthopaedic Surgeons in San Francisco, Dr. Clive Solomons of the University of Colorado Medical Center told of a study of 30 children with this rare disease. Seventeen of them seemed to have improved while taking magnesium oxide tablets. These 17 had one to three fewer fractures each during the 18-month trial period than in a prior 21-month control period. Ten of the 30 showed no change, and three had more fractures, but most of the patients also had a significant increase in bone density (necessary to prevent fractures) and those with fewer fractures sweated less, and were more active.

Dr. Solomons and his associates tried magnesium oxide (MgO) because studies had shown it protects part of the bone against thyroid hormone, which antagonizes a substance called calcitronin and then causes an increase of the substance that inhibits calcification. He said that magnesium also inhibits secretion of parathyroid hormone which may help maintain positive calcium balance, of which there is not enough in brittle bone disease.

The researchers also found what they believe may be two signs of subclinical osteogenesis imperfecta—abnormal platelet aggregation and leukocyte metabolism. Platelet aggregation tests of three patients and of two parents who were presumed asymptomatic carriers turned normal after each took MgO for ten to 16 months. In 14 patients and 22 asymptomatic relatives, white cells reduced a specific dye better than it did in controls—a measure of the efficacy of the treatment.

If you are the parent of a child with this rare disease, the only way to keep him or her from repeated fractures is to practically sandbag him in bed—keeping pillows around both sides as well as at his head and feet, and not permitting any heavy toys or books to be used by him.

You might also like to read *Give Every Day A Chance* by Beverly Plummer, published in 1970 by G. P. Putnam's Sons, about Mrs. Plummer's daughter, Roxie. After many

weary years of going from one doctor to another, one hospital to another and suffering through the child's many fractures and illnesses, Mrs. Plummer is now able to report that Roxie drives her own car, goes to college, and seems well on her way to a normal adult life.

We are certain that newer and improved research will find a way to help your child with this rare genetic disease.

Congenital Eye Problems: Besides the rubella-induced eye problems already mentioned, there are physical anomalies of the eye reported upon by the June 15, 1970, issue of *Modern Medicine*, in its "Lecture Series" section, that may sound strange, unless you see one or more of these problems.

An encephalocele is a herniation between the eyes because of a defect in orbit-presenting at the upper root of the nose. Another kind is an intraorbital herniation causing proptosis and downward displacement of the eye globe. These abnormalities, and similar ones which are part of a developmental abnormality in which intracranial structures herniate through a bony defect in the orbit usually above and medially, are usually present at birth. The eye may be displaced and ocular rotation impaired as a result of pressure upon the eye from the mass and surgery must be done by a neurosurgeon to effect immediate repair.

Dermoid tumors are congenital and may be located on the eyeball, conjunctiva, lids, or orbit. The most common location is on the orbital rim. Dermoids may appear as cystic or solid masses and are usually a benign type of tumor.

Hemangioma is the most common benign orbital tumor of childhood and usually becomes apparent shortly after birth. It grows rapidly in size during the first few months of life, the skin over the eyelids may develop a purplish color, due to an underlying vascular tumor. It will either undergo spontaneous remission or it can be treated with X-rays. It tends to be diffuse in children, but is localized in adults, at which time it can be removed by surgery, according to the *Modern Medicine* article.

Glioma of the optic nerve is an occasional cause of eye problems in children and appears during the first decade of life. Impairment of visual acuity is a problem because the eyeball is displaced directly forward. The glioma must be removed surgically, but if the damage to sight has already occurred, there is no cure for it.

Many ocular infections take place during childhood. A gon-

ococcal infection of the birth canal of the mother can cause ophthalmia neonatorum in the infant. Signs of this ocular inflammation occur within 48 hours of birth and if not treated immediately and completely, permanent corneal damage to the baby's eyes may occur.

Ocular vaccinia is incurred through inoculation, from the child's own vaccination or that of another child's vaccination site. Ulcerated lesions, usually unilateral, may appear on the eyelid, conjunctiva, or cornea. Keratitis may threaten vision, as the conjunctiva is inflamed and may show grayish ulcers. Usually local antibiotics are given to prevent secondary infection.

Chicken pox may cause lesions similar to those of vaccinia. Photophobia, tearing, and redness indicate ocular involvement and prompt therapy is required.

Acute catarrhal conjunctivitis is a common childhood disease. The conjunctiva is markedly injected and a bad secretion is present on the lid margins and eyelashes. It may be bacterial or viral. Severe pain and photophobia suggest the possibility of a secondary keratitis, but this usually subsides promptly with local antibiotics.

As they look at their newborn infants, parents often wonder what the baby sees when he opens his eyes. Does he see his parents' faces? Does he notice differences in brightness or color? Although these and related questions have long intrigued scientists, valid answers have been slow in coming because of the obvious technical problems associated with research on newborn infants.

At Cornell University a number of studies in the past few years have shed light on the development of vision in the first few months of life. Back in March 1970, some findings were announced by project leader John Doris, associate professor of human development and family studies in the New York State College of Human Ecology, Cornell.

"Scientists know from research in recent years," he pointed out, "that although an infant has some visual function within a few hours after birth, the efficiency of his eyes is quite limited. We know that the newborn infant's visual acuity—the ability to distinguish fine details—is poor. In fact, it would be no better than 20-400 on a Snellen eye chart, in comparison to the 20-20 vision of a normal adult."

The Cornell studies, which involved the carefully controlled observation of infants' eye movements as they are

shown a series of contrasting gray stripes, have been primarily concerned with brightness-discrimination and its relationship to the ability to distinguish fine details. The research has demonstrated that brightness-discrimination undergoes rapid development in the first two months of life. It also found that the average newborn baby is unable to respond to anything other than a marked contrast in brightness, whereas the two-month-old child is able to discriminate subtle differences in contrasting shades of gray.

The retina—the light sensing surface at the back of the eye—is incompletely developed at birth and consists primarily of rods, the sensory elements that are the most active in night or twilight vision. However, during the first two months of life there is rapid development of the cones, which are the elements responsible for daylight and color vision, Doris noted. Of particular importance during this time is the refinement of the cones in the fovea, which is located in the center of the retina. These cones are responsible for the development of the ability to perceive fine details, the researcher said. Doris, who specializes in clinical child psychology, is co-author of a recent book, *Psychological Problems in Mental Deficiency*.

Another researcher, Mitchel Title, M.D., who is a New York ophthalmologist, warns in a recent issue of *Sandoz Panorama*, a medical publication for doctors published by Sandoz Pharmaceuticals in New Jersey, that the mistaken notion that a child's eyes cannot be examined until he has learned to read may lead to irreparable harm to the child's eyesight. According to Dr. Title, a child's eyesight should be checked as regularly as growth, weight, and neuromuscular development. If eyesight is ignored until school age, it is frequently impossible by therapy to modify abnormalities of visual development. Important eye problems affect almost ten per cent of all children under the age of seven.

The ophthalmologist pointed out that the stages of visual development should be "observed earlier and more frequently than is done at present." He believes the pediatrician or the family doctor who treats young children should familiarize, and equip himself with the few basic techniques necessary to screen children for ocular abnormalities. These include evaluation of the child for the "normal attainment of milestones of visual development, evaluation of visual activity, evaluation of muscle balance, and evaluation of the clar-

ity of the optical media." During the first six months of life, both physicians and parents should watch for reaction to light, or random eye movements which may represent poor bilateral visions. The physician, by covering each eye in turn in the presence of an attractive toy, can check for unilaterally impaired vision. "The infant who does not object to having one eye covered, but yowls and squirms when the other is covered, is merely indicating his frustration at no longer being able to see the toy and therefore is telling us of his unilaterally impaired vision."

By the age of three, Dr. Title believes, most children can be tested for visual acuity through the use of the "E" chart, in which "E's" of different sizes are pointed in various directions. He feels too little attention is paid to early eye conditions and serious, permanent harm can result from such neglect.

Hearing Problems: At least one out of 100 babies born to a pregnant woman who suffered from rubella will either be born deaf or have a serious hearing defect. Dr. Erik Wedenberg and Dr. Bjorn Westin, both of Stockholm's Sabbatsberg Hospital, believe that the earlier a defect is detected, the more can be done to beat the handicap. The first year and a half, according to Dr. Wedenberg, may be the most audiologically important time in the infant's life.

"The baby produces sound all the time," says Dr. Wedenberg, "listens to other sound sources and tries to imitate them. He needs this auditory feedback. If this stops, he stops making sounds and retreats into a silent world."

Family Health magazine, in its August, 1970, issue, told of the four-month-old baby that did not stop crying when his mother came into the room, even though she spoke soothing words. But when he caught sight of her, he calmed down. Similar occasions caused the worried mother to wonder if her child had a hearing problem. A pediatrician confirmed her suspicion.

"All too often, hearing loss in children is first noticed by the parents," the *Family Health* article pointed out. "Naturally, pediatricians and ear specialists would like hearing defects to be detected as soon as possible, so that special training can be started from infancy.

"Two California physicians have now devised a simple hearing test that hospital personnel can administer to newborn infants," the article continued. "Dr. Rayford C. Reddell

and Dr. Donald R. Calvert developed the technique in the course of a three-year investigation at the San Francisco Hearing and Speech Center. A day-old baby, disturbed by the noise from the amplifier held over his head, wakes up crying, proving his hearing is normal. Other typical reactions are jerking, blinking, and change of breathing rate. A deaf child does not react.

"Until hospitals and physicians give such tests routinely, parents must continue to pick up the first clues. New York pediatrician, Dr. David Kligler of Albert Einstein College of Medicine, says: 'When his mother enters the room, a one-month-old baby with normal hearing usually quiets down even before he can see her.' By nine months of age the child should react to the word 'no,' and by 15 months follow a command to get a toy."

Aphasia is an impairment between hearing and connecting what is being said, and is a condition something like hearing a foreign language for the first time. You hear it, the sound, but you don't know what it means. This is not deafness in the true sense, nor is it retardation, lack of intelligence, or a form of mental disturbance. Actually, aphasia is caused by a neurological malfunction of the brain and needs to be helped by special speech and-or remedial reading classes.

Audiologists and otologists prefer to call all forms of deafness in children by the term "profound childhood deafness" and admit that while some types of birth defects in hearing are caused by genetic autosomal recessive tendencies, these develop primarily through family inbreeding and the fact that too many deaf people tend to intermarry with other deaf people, giving the genetic fault no chance to be eliminated by marriage with normal hearing people.

Many of the birth defects we have already mentioned include hearing deficits among their damage, and some researchers suspect viruses other than that in maternal rubella with causing the fetus to bumble badly when it comes to the development of the ears and hearing. Jaundice, erythroblastosis, and other factors are also suspected in cases of infantile deafness. Central nervous injuries can also affect the ears, and shocks of any kind either in the womb or after birth may harm the ears or nerves that send messages to the brain.

A baby may be born with seemingly good hearing and then when only a few months old get a high fever or an inner ear infection that will leave hearing problems. How-

ever, if these infections are caught quickly enough, they do not advance to the point where an eardrum must be opened or a mastoid operated upon. It is also wise to be concerned when a child with a cold or any kind of fever or infection pulls at the side of his head or ear. This is an indication that the infection may have landed in the ear's eustachian tube or some other area where it is important to have prompt treatment.

On February 22, 1970, science writer Dolores Frederick of the *Pittsburgh Press* revealed a new breakthrough in surgical treatment of the ear. She even revealed something few people usually know—that everyone has two ear drums. You can't see them, but they're so tissue-thin inside the ear that they vibrate with the slightest sound.

"Three years ago," the story went, "there was little hope that ear drums could be transplanted. Then a Belgian surgeon perfected a technique and performed the operations on hundreds of patients. . . .

"The drum normally curves into the head—like a contact lens—but after transplant surgery, it sometimes was healing as a flat disc and patients couldn't hear as well as they would if it was curved.

"But Dr. Ralph J. Caparosa of Eye and Ear Hospital (in Pittsburgh) found a way to help the transplant heal without 'shrinking.' He 'reinforces' the transplanted drum with a piece of tough fiber—called fascia—taken from the back of the patient's scalp. Since he perfected the technique (in 1968) he has performed ten such operations and he said the results have been very good. If the drum isn't completely ruined, he covers the hole . . . with a piece of scalp fiber. . . ."

The operation takes about two hours and must be performed under a microscope because of the tiny bones and ear canals. A course to keep doctors abreast of ear surgery and correcting hearing problems is given three times a year at the Pittsburgh Eye and Ear Hospital, cosponsored by the University of Pittsburgh.

Miss Frederick's story concluded: "Broken ear drums sometimes appear in patients who may have had scarlet fever in their younger years. Ear infections and scuba diving accidents are other causes. But Dr. Caparosa foresees a drop in the number of cases because . . . antibiotics . . . have cut down on ear infections."

3

Living More Easily with the Birth-Defect Child

There are four specific factors that enter the lives of a family with a birth-defect child and must be lived with as long as that child needs help. They are: education, medical care, home programs, and recreation.

This book is concerned with the child until the primary school age—usually around six or seven years of age. For further help, we shall give you other books and groups to consult and ideas later in this volume. Some of the most successful ideas given here, however, are examples of what families, volunteers, communications, organizations, and individuals are accomplishing by working together or just with the child.

Unless a child is so severely handicapped mentally or physically that he cannot be cared for at home, and must be institutionalized, the very best thing for him is to be raised with his own family, to be given love, and to never be allowed to consider himself a burden. Naturally, he will require more attention, service, and concern than a normal child, but at the same time he will return to you lessons in patience, humility, spirituality, and love that you would otherwise not have known, and these are the kinds of lessons we all require at some time or other for the conditioning of our own hearts and souls.

No matter how handicapped a child, he must be taught to try to meet the same challenges that other children meet, and to accept the same punishments when he refuses to do his best or if, somehow, he lets his handicap hurt others unnecessarily. It is frequently very difficult for a parent or other relative to scold or punish a child when he needs it, particularly if that child is smart enough to use his defect or illness as a way of punishing the family member.

Parents and other family members living with a birth-

defect child should try to have family counselling in a group or by an individual who is an expert in children with handicaps. Such counselling will tell how to cope with the day-to-day behavior problems that may arise. It can even be your pediatrician or a mental health expert, perhaps someone with the volunteer organization concerned with the birth defect your child has, or perhaps a friend with that kind of knowledge, who can give you this type of counselling.

Never try to make your own behavior rules for your birth-defect child. You will find they will frequently be the kind to ease your own conscience, rather than to strengthen the child's character, and sick or well, children do have their characters that must be considered and individually shaped.

This writer learned much of this during her adopted daughter's childhood with the innumerable infections that resulted from the lack of antibodies in her blood. During Claudia's high school years, much of her education was at home, because she had to be in bed so much that she attended the public school too infrequently. New York City has a visiting teacher's program for homebound children, and perhaps your area has one, too. No matter how sick our daughter said she felt, she was never allowed to disappoint this home teacher, who only visited two or three times a week, because Claudia could always read and she could always write, and she had no other commitments. However, she much preferred being coddled and watching television, or she would miraculously become better for the day she knew we hoped to take her to a party or visiting someone's home.

Claudia lost the use of her TV set for a week each time she disobeyed our rule not to watch it until her school work was finished. She lost a birthday party of her own that was to be held in a restaurant, for not getting good marks from her teacher that month. She also lost the privilege of having friends come in to play whenever she pulled unnecessary tantrums during their visits. We allowed her the normal quarrels of youngsters, but she was not permitted to feel that because of her illness, nobody else could touch her toys or books, or listen to her recordings. She had to learn that sharing was a privilege and to share whatever she had, including smiles.

When she had the energy (and this was her doctor's decision, not ours) she was expected to dust her room and put

away her own things. Even in bed, before lights went out, everything she had used that day must be removed from her bed and at least piled up neatly on the chair, ready for use the next day. If she refused to do this, everything on the bed went into a drawer while she was asleep, and until she was well enough to get out of bed (again with the doctor's permission) she could not use any of those things again, unless they were her school books and work.

The times when she had high fevers and was only partially conscious were the times of my own conscience and worrying that I might have been too harsh with her. But I had good medical friends, and good psychologists to advise me, and I appreciated their warning me that the greatest danger to a child with a long-term illness or handicap was being made to feel rules were not for them to follow. In their secret hearts, sick children like to feel like "one of the family" most of the time, they like having special responsibilities, even if it's having a smaller child (a new baby perhaps, or a visiting one) put in his carriage or bassinet nearby so the invalid can "baby-sit" and feel important with that special duty.

The handicapped child who is permitted to chop up food for mother, or be put in a special wheelchair near the sink to do the dishes, or to regulate the washing machine and possibly take out the clean clothes and put them in a basket—no matter how long it takes him, whether he has a palsy and drops things, no matter what his defect, let him know there is something he can do at home so that he can be told "well done, dear" many times, just like the other family members.

If your birth-defect child has a special talent such as singing, writing, creating music—anything which can be utilized in community programs, special events or perhaps can help to raise money for something the family or the community is doing—he should be asked to contribute this talent to their efforts. At the same time, no more should be made of his having such talents than would be done for others in the family having the same talents. The point to make is not that he is special in having a talent despite being sick, but that he is special only because he is making good use of that talent for others.

If you use television as a "baby-sitter" when the child is old enough to talk and report impressions, you should make certain that he knows what he is watching, and discuss it

with him, have him tell you the stories he watches, ask him to tell you characters that were portrayed, and let him give you his impressions of whether or not he likes something or agrees with it.

Do not let programs like "Sesame Street" be your child's tutor. They are aimed primarily at "entertaining," regardless of their promotional pitch, and many educational experts disagree that they actually teach very much. If your child's handicap requires exercise, find programs that may help him with this. Try to do those exercises with him, either using the televised instructions, or music on his own little phonograph. Of course, there will also be the special exercises medically prescribed for him, and these can seldom be done alone by him, or even with another child. It is always wisest for one adult to join the ritual.

Toddlers and growing children are not automatically inspired to help themselves, but your other children can each become a part of the program for the handicapped one, using games and music and other outlets for their participation, instead of the medically-oriented ones. It is always good to have children in on family routines for the birth-defect child, not as a baby-sitter but as someone else the handicapped child can live with, react to, and grow with.

If the birth-defect child is your only one, encourage the older children of your other relatives or neighbors to come by frequently. Perhaps you can have skating or apple-bobbing parties in the park, or even drawing sketches at home for an hour or so. Even when your own child may not be able to participate effectively, he can keep score, be allowed to count out loud with the others, or in some way be made to feel he is part of the "game."

When you must visit clinics with him, be sure to always have something along that will interest him or something that he will be able to share with another child his own age while you wait. Most clincis for birth-defect children these days have special recreation and waiting rooms, many of them with toys or books, but again, your child may prefer his own and if he is an only child, this can be an excellent sharing experience for him with another child.

Try not to lessen his feeling of progress health-wise by allowing older family members (like grandmothers and aunts) to weep over him or fuss over him and make him feel like someone who is a vegetable. When there is family grief over

someone or something else, let him share that grief and be told the reasons, but try not to express grief over his own condition, because then you will make him feel he is going to die, is unwanted, or perhaps it will give him a suicidal complex as he grows older but does not seem to get better. Nobody wants to be considered a burden, and your own problems will be less when you stop reminding the child (and yourself) about his handicap. You should not allow others to do this to him, either, no matter how well-meant their remarks and excesses.

Let us say your child has a very rare birth defect like osteogenisis imperfecta (brittle bone disease) and in your area you know of nobody else having a child with this problem. For yourself, rather than your child, it is important to know another family facing the same things you do, because you need to know how they cope. The way to learn quickly whether there *is* another case as rare as your child's is to call or write the city editor of your local newspaper. Uusually, such an editor will want to do the story, and will send a reporter and photographer to your home, and shortly after the story appears you will hear from those who know someone with such a condition.

Or perhaps your doctor will check with other doctors in the hospitals where he works. Or you might contact the County Medical Society in your area, and ask them to let you know whether any of their orthopedists (or the kind of doctor who handles the kind of disease you might be looking for) have such a case near by.

The relief of knowing just one other family that is coping successfully with what has until then been something you could share only with your doctors, your family, and a few special neighbors and friends is very real indeed. We never found anyone with the strange condition our Claudia had, and because she was adopted we had no genetic background to help us diagnose her condition quickly. She nearly died a dozen times and it was bitter indeed to have a great many people—including medical ones—decide out of clear air that there was nothing really wrong with her besides something "psychological"—that it was probably induced by my own "over-protectiveness" and other such rot. How many times I asked myself whether I was being "over-protective" when I sat up all night putting ice-packs on her, rubbing her down with a mixture of alcohol and water, giving her medications

every two hours, and repeating this routine day and night until she got better.

How many times she was rushed to the hospital at all hours. Once, a doctor carried her in his arms (she was 13 and already weighed much more than she should) because her two ears were so badly infected that she screamed with pain at laying her head flat.

My only reason for including these stories about Claudia's problems (and letting you know that eventually a smart hematologist found out that it was a blood problem that a repeated series of gamma globulin injections eventually brought back to normal) is to advise that you must not let even doctors' opinions make *you* feel to blame, unless the doctor is a psychiatrist who has been recommended by your own doctor to talk with *you*. One neurologist looking Claudia over was also a psychiatrist who told the then 13-year-old child there was nothing wrong with her that some analytical sessions with him would not cure. You may be certain that when she told this to me, the doctor was immediately discharged and a letter sent to his County Medical Society for his unethical conduct. No child should be given a diagnosis—only the parents, and this is very important for you to remember.

Do not take advice, blame, or even praise from anyone who is not authorized to help you professionally with your child. You will need professional help in everything, and discard the efforts of do-gooder amateurs to advise you. No matter how much they want to help, nobody can advise you on whether you are doing the right or wrong thing for your child except other parents with children having the same condition as your child, and-or your doctors and therapists who are helping your child.

However, you must recognize that the mothers and family of other birth-defect children cannot be considered amateurs, either, since they have probably been working as volunteers in the organization helping such children even longer than you may have. Sharing ideas and experiences with such people is probably better therapy for you—and your marriage partner—than a vacation. You will then realize that others have come through this situation in spite of all the fears and problems that the first knowledge of your child's condition brought to pass.

Professional and Volunteer Organizations:

Blood Diseases: The Children's Blood Foundation, Inc., 342 Madison Ave., New York, New York 10017, (212) MU-7-1564. This is not the hospital, but the office of the organization, and they cooperate with parents and blood groups throughout the United States, advising the location of other foundations near you, or advising doctors on treatment for specific diseases. The New York City hospital group takes patients from both New York and New Jersey, but makes referrals for other areas.

Neurological Problems, such as Spina Bifida: The National Foundation-March of Dimes Birth Defect Center nearest you will advise whether or not they handle this kind of problem, or will direct you to the nearest one that does. As an example of what such a center can do, in New York Hospital, New York City, Department of Pediatrics, the meningomyelocele (spina bifida) program takes care of 100 children with this or any other developmental anomaly of the spine as well as hydrocephalus, in a new clinic established under the direction of Dr. Alfred L. Scherzer.

The program staff includes pediatric interns and residents; two nurses (one with public health training); one social service worker, and one psychologist. Direct consultation services are available from the departments of neurology, urology, orthopedics, neurosurgery, and other specialties as needed.

Prior to the actual clinic session on Thursday afternoons, a discussion group is held for one hour with parents whose children are scheduled to be seen. These meetings provide an opportunity for exchange of information and ideas regarding management as well as counselling and support. During these sessions, the children are cared for in the pediatric playroom and for many this is the first opportunity to socialize with children having a similar handicap. Nurses and social workers meet with parents prior to the medical examination to review care of the child and special problems of management.

One nurse specially trained in the care of the ileal conduit bag (used because of urinary problems coincident to severe neurological conditions) reviews the appliance and makes recommendations for any change. A physical therapist teaches parents special exercises to be done at home. Public health nurses, social service workers, and a staff from other community agencies working with patients attend special ses-

sions for discussions of patient needs. The charge is minimal.

The aim of such a program (and similar ones are held in hospitals and clinics elsewhere for other handicaps and birth defect problems) is to bring together more closely the medical specialties involved, improve coordination of the variety of outpatient department services provided and to develop closer relationships with pertinent community agencies. Such a new form of comprehensive care is serving as a model for medical management of any pediatric chronic disease condition involving a variety of hospital specialties and services, and can ultimately be extended to other clinics of the pediatrics department.

Cerebral Palsy Programs: The national office of this handicapping problem is United Cerebral Palsy Associations, Inc., 66 East 34th Street, New York, New York 10001, (212) 889-6655. The main New York State office is at 220 West 42nd Street, New York, New York 10036, (212) 564-2460, while the main New York City office is at 339 East 44th Street, New York, New York, (212) 661-0900.

You may write to the national office for the location of the nearest branch of their organization in your town or city. Most cerebral palsy groups are staffed by both professionals and volunteers, and usually include a treatment center, either one of their own or affiliated with a hospital or medical complex near by. For instance, in Rochester, New York, the Cerebral Palsy Treatment Center of East Avenue is owned and operated by the United Cerebral Palsy Association of Rochester. It is largely supported by Community Chest funds. Children are accepted for treatment and training if:

(1) they have cerebral palsy,
(2) they are able to benefit from the treatment, training, and educational programs offered by the Center,
(3) there is no other treatment or educational facility that can meet their needs better, and
(4) they live within traveling distance from the Clinic.

Children are recommended for treatment and training by the Cerebral Palsy Diagnostic Clinic after a thorough examination by a team of specialists. Various types of treatment are offered including physical, occupational, and speech therapy. There is also a nursery school for the development of socialization of the young children. There is a first grade for

the older educable children who are not yet ready for the classes for the orthopedically handicapped children. There is also a pre-nursery school group to meet some of the needs of the very young cerebral palsied child and his parents.

To keep the parents informed about their child's progress, a parent day is held monthly, usually on Friday afternoon. At this time, the clinic program is curtailed so that each teacher and therapist can spend as much time as possible with the visiting parents to discuss what is being done for the child at the clinic and how these efforts can be carried on in the child's everyday living at home. The parent-day program gives the staff and parents an opportunity to get to know each other and to learn what each is doing to help the child make the most of his abilities.

Evening seminars are offered to acquaint parents with basic concepts of cerebral palsy such as what causes it, types of treatment and problems of training, education, and general care. The parents' participation enables many ideas and experiences to be exchanged regarding some of the problems arising in the care of cerebral palsied children.

To facilitate the child's attendance at the clinic, a transportation system has been established which takes the child from his home to the clinic and back. A physician is regularly on call for any emergencies which may arise. There are regular orthopedic and psychological examinations at the clinic on which are based changes in treatment and education. An eye specialist examines those children with eye problems when necessary. Other medical consultants and agencies are used for help as necessary. The clinic's social worker is always available to discuss problems about which the parents are concerned.

Cerebral palsy is due to incomplete development of, or damage to, parts of the brain which control muscle function. This developmental defect or damage may be limited to the motor apparatus or may involve parts of the brain which have other functions. Cerebral palsy, therefore, may be associated with handicaps other than the difficulty in muscular control.

A common physical problem occurring in 50 per cent of cerebral palsied children is turning in or turning out of the eyes. This is sometimes called strabismus or squint. It is due to an imbalance of the muscles which control the eye movements in most instances, but is reparable by surgery in which

these muscles are lengthened or shortened to bring the eyes into proper alignment, resulting in both better vision and appearance.

Another visual difficulty which occurs in a very small proportion of children with brain damage is reduced vision due to damage of the optic nerves or the visual tracts of the brain. This condition, sometimes called optic atrophy, results in partial blindness and is not reparable by surgery or glasses.

Hearing difficulties are not uncommon in children with cerebral palsy. It is said that about one-third of athetoid children have some degree of hearing impairment. There may be partial or complete deafness. The hearing ability may be reduced at all pitch levels or reduced only for certain pitches. The latter hearing defects are referred to as "tonal gaps." Children learn to speak by repeating what they hear; those with tonal gaps—that is, those who hear some sounds well and others not at all—hear a distortion of the human voice and reproduce these distorted sounds; this speech is difficult to understand. The speech pattern may be further complicated by the incoordination of vocal control due to athetosis or slow speech development. Children with hearing defects, therefore, should have a speech and hearing evaulation. A speech therapist can be consulted to help the parents to understand the child's problem in speech development.

Some cerebral palsied children have a reduction in the sensation of pain, temperature, and touch. Knowledge of this is useful in helping the child in occupational therapy where hand and arm use are developed, since hand use is dependent in part upon sensations from the fingers and hands to the brain.

Convulsions are not uncommon with fever in children without cerebral palsy, and they occur more frequently in cerebral palsied children, with or without fever. Seizures are usually not dangerous; they last from a few seconds to ten or 15 minutes. If the child has a fever, cooling him down by removing his clothing and sponging him with lukewarm water before a warm oven usually suffices. If the convulsion occurs without the fever, the child's clothing should be loosened, a pillow should be put under his head so he won't hurt himself, and a wad of cardboard or piece of soft wood should be placed between his teeth to prevent tongue biting. Most children with cerebral palsy do not bite their tongue during a convulsion, but occasionally this does occur. A phy-

sician should be called in accordance with the parent's experience and understanding of the condition. Most convulsions can be controlled by medication.

About half of the cerebral palsied children have a significant degree of mental retardation. These children require a specialized educational program which attempts to develop their abilities to learn. Since there are various degrees of mental retardation and associated learning difficulties among these children, there is need for individualized training and education by competent, specially trained therapists and teachers. Mental retardation does not imply that the child cannot learn, but that his rate of learning is slower than that of the average child, and the development of his learning capacities will be limited. Care should be taken not to attempt to increase the child's intellectual abilities through more intensive training and education since this practice may defeat its aim by developing resistance to learning.

Because of the muscular imbalances and, particularly, because of spasticity in the muscles, orthopedic deformities are prone to develop. Wild dislocations of the hip, toe-walking, turning in of the ankles, tendencies to cross the legs and postural difficulties are not uncommon in cerebral palsied children. The use of pillow-pants to prevent dislocations of the hips by aiding the development of deeper hip sockets, "skis" on the shoes to lengthen the heel cords so the child can walk with the flat of his foot on the floor instead of on his toes, bracing, casting, and special shoes are some of the measures used to prevent the development of orthopedic deformities.

Cerebral palsied children have certain other problems in common. Here are a few from the Rochester group, whose bulletin has supplied all of the cerebral palsy information in this section:

Feeding Problems—Because of the involvement of the tongue and jaw muscles, the cerebral palsied child has difficulty chewing and swallowing, particularly the younger child, who may require a liquid diet for many months. Soft and chopped foods help the older child who cannot chew well. If the tongue is weak or poorly coordinated, it may be necessary to tilt the child's head backward so the food will move by gravity to the back of the mouth where it can be swallowed. Care must be taken to avoid choking, which frightens children.

Constipation—This common symptom occurs mostly in the

young cerebral palsied child and improves with age. Conservative remedies such as prune juice and other laxative fruits or their juices are best. When diet does not control the constipation, milk of magnesia is usually effective. If this fails, a physician should be consulted. When the child becomes more active, this problem improves and eventually disappears.

Startle—Most cerebral palsied children have a persistent startle reflex, that is, loud noises or the sudden occurrence of the unexpected, make them momentarily excited. All babies are born with this tendency, but cerebral palsied children continue to startle for an indefinite period. It is related to the brain damage, and usually improves as the child grows older, often disappearing before five years of age. Because of this tendency, sudden loud noises and other surprises should be minimized, until the child can handle them comfortably. Some children are also sensitive to high pitched sounds like sirens and cover their ears or cry when they hear them. This, also, seems to be related to the brain damage and improves as the child matures.

Fear of Falling—This is a common fear of children who have difficulty in maintaining balance. It is worse in tense, insecure children. It often seems to be related to their realization that they might fall and not be able to protect themselves. The best treatment is to make standing up and moving through the air fun, so they can learn to be comfortable and find pleasure in being off the ground. Children who have the least fear of heights or of falling have learned to control these fears in games with their parents who were careful not to let them be hurt.

Surgery for Cerebral Palsied Children:

About 100 years ago, Dr. John Little, an orthopedically handicapped person, cut the heel cords of persons with muscular contractures of the calves of their legs so they could stand in a more normal position with their feet on the floor instead of the toe-down position. Since that time, orthopedic surgery and its allied therapies have continued to be the most important therapeutic tools in the treatment of the musculoskeletal disabilities of persons with cerebral palsy.

Cerebral palsy is due to either lack of development or damage to part of the brain which controls the tone and co-

ordination of muscles. In the spastic, the muscle tone is increased but the muscle strength is weak. In the athetoid, there is difficulty in coordinating muscular movements; the muscle strength is good and the muscle tone may be normal, but when the individual is under tension, the muscle tone may increase.

The ataxic tends to be weak due to reduced muscular tone and has poor muscular coordination. As he matures and gains in strength and experience, the coordination tends to improve. Experience has demonstrated that treatment of the physical handicaps of cerebral palsy should be directed toward:

(1) teaching the child to make the best use of those abilities he has,

(2) preventing the development of permanent contractures of muscles and the resulting bony deformities through mechanical devices such as pillow-pants and "skis",

(3) correcting the muscular imbalances, and the resulting bony deformities and nonfunctional positions of the extremities to make it easier for the child to use his muscles in a way which will be of greatest advantage to him.

Speech therapy is directed toward improving the child's word formation through teaching him to make vocal sounds which are difficult because of poor tongue control or irregular breathing.

Occupational therapy focuses on improving the use of the arms and hands through training and attempts to prevent and correct contractures which impair the best use of the upper part of the body.

Physical therapy attempts to strengthen the muscles of the back and legs and, through training, increase their function in sitting, standing, creeping, and walking.

In the cerebral palsy treatment program, the philosophy of treatment is to motivate the child to actively participate in the training exercises in a way that hopefully will carry over in his everyday living. This "kinetic" treatment develops muscular strength and abilities as the child is taught new skills which will be useful to him.

The role of orthopedic surgery is to minimize the effect of shortened muscles due to permanent contractures which

cause the legs to remain partially crossed (scissoring), the hips and lower legs to be flexed, and the foot to remain in a toe-down position. By lengthening tendons such as the heel cords, transplanting the other attachments of muscles to the bones of the body and correcting such skeletal defects as persistently dislocated hips and unstable feet and ankles, orthopedic surgery increases the mechanical advantage of the child's muscles so that greater functional use is possible. Orthopedic surgery, therefore, does not cure cerebral palsy, but does decrease the muscular and skeletal disability that may result from contractures of the muscles.

Skill and experience are necessary to decide which child might benefit from surgery. Sometimes the aim of the surgeon will be limited to improving the child's sitting comfort. With the more educable child who has more muscular strength and can be motivated, walking may depend upon corrective surgery.

When a child goes to the hospital for a surgical procedure, he should have some understanding of where he is going, if he is old enough to understand language and explanation. The details of the operation need not be discussed with him, since he will be asleep when it is done. It is more important to give him an idea of where he is going and to assure him that he will return home from the hospital. When a child is hospitalized for surgery, his morale is improved by regular visiting by his parents and those he loves, and if the mother can arrange to be there all day and evening on the day of the operation, so she can comfort and help him when he awakens from the anesthesia, that will be even more helpful to his morale.

Surgery is usually followed by physical and occupational therapy to capitalize on the therapeutic results of the operation.

Other Ideas on the Handicapped:

The New York University Medical Center's Institute of Rehabilitation Medicine headed by Dr. Howard Rusk, the rehabilitation expert, announced in July, 1971, that it was pioneering research efforts directed toward the very young, severely handicapped children—infants 18 months to three years—to help them learn to adjust to their physical limita-

tions and grow toward emotional, social, and educational maturity.

The infants live in a four-bed nursery at the Institute, a "home" featuring kiddie-sized furniture and specially modified bathroom and playroom facilities. During an intensive two to three month program they are visited often, even daily, by their families who participate in the therapy programs. When their children return home, the families will continue these programs under the guidance of a home visiting preschool specialist.

In the nursery, the infants receive therapy—education, physical, occupational and speech—according to their individual needs. They also are taught self-feeding and personal grooming, as well as how to live with others and accept themselves as worthwhile human beings.

Another innovation at the Institute is the new outdoor playground for exceptional children, both the wheelchair-bound and ambulatory ones, as well as those with and without limbs. It is located in mid-Manhattan, a couple of blocks from New York University Hospital, on a 120 x 30 foot plot, and was financed through a $75,000 grant from the Bureau of Education for the Handicapped of the U.S. Department of Health, Education and Welfare. It took the architects and therapists ten years of joint research before the project was launched, to make certain it would fit all the needs of those for whom it was intended.

Among its unique facilities are rubber-edged sand-tables with circular cut-outs to accommodate wheelchairs; a playground floor made of either grass or a green synthetic rubber compound hard enough to accommodate crutches or wheelchairs, but soft enough should someone fall; a tree house that can be reached by ramp or ladder; a waterfall which empties onto tables for water play with plastic toys floating therein; a periscope to view the world outside the playground, and a number of other educational toys, such as scales and pulleys, as well as numerous handrails to make certain of the security for each child's movement.

The director of the project is Mrs. Ronnie Gordon, an assistant professor of clinical rehabilitation medicine at the Institute who is in charge of all preschool developmental programs there. "The children learn how to adjust themselves to a ramp and can balance on a mat, as they learn to share with

a child who wants to use the same piece of equipment," she explained.

Because the playground is the first of its kind for multiply-handicapped children, it serves not only the children of the Institute but also all other exceptional children in the community. "We would hope that this demonstration playground will prompt the New York City Department of Parks to consider providing similar little play areas as appendages to existing children's parks throughout the city," Mrs. Gordon announced. "We must be concerned with stimulating the development of all children."

Of course, there is nothing to stop other groups in other cities from visiting the playground, consulting with Mrs. Gordon, and applying to their federal and state bureaus for education of the handicapped for funds to establish similar projects for the exceptional children in their own areas.

When it comes to educating handicapped children, an interesting three-year project that runs through 1972 has been operating jointly under the direction of the University of North Carolina and the University of Arizona. The U. S. Office of Education gave a $288,000 grant for that period, and it is hoped the grant will be extended for another three years.

The initial goal was to train regular classroom teachers to work effectively with children who "suffer handicaps, whether physical or emotional," and the next step was the development of a technique to bring the handicapped child back into the regular classrooms or to keep him from being transferred out of the mainstream of education.

In North Carolina, 20 teachers work each summer at the Wright School for Handicapped Children in Durham, and go back to their regular classrooms in the fall with their special knowledge. In Arizona, ten Indian teachers are trained at Camp Echo, a summer facility in Tucson, Arizona, for physically handicapped children. The children involved included both emotional and physically handicapped children from the minority groups, and the project attempts to increase the behavioral, social, and scholastic competence of such children, at the same time helping to produce more effective, understanding, and skillful teachers.

Another summer project that could well be a pilot example for other areas is the Riley Memorial Association Camps for Physically Handicapped Children which in July, 1971,

enjoyed its 17th season of outdoor camping at Camp Riley in Bradford Woods, Indiana under sponsorship of the James Whitcomb Riley Memorial Association.

According to the *Journal of the Indiana State Medical Association*, the camp program, begun in 1954, provides a unique and beneficial experience for some 180 crippled and handicapped children who, because of their disabilities, have few opportunities for outdoor living, education, or adventure. Any physically handicapped child from eight through 15 years of age is eligible to apply. His own physician must sign the application after which a committee of doctors, nurses, and medical social workers at Riley Hospital determines whether the applicant can participate in and benefit from the camp activities. Children in wheelchairs, on crutches and braces, and a limited number of deaf and blind children are accepted each year. The camp can accomodate about 70 campers for each session. The first session begins July 12 and ends July 24; the second runs from July 26 to August 7 and the third and final session opens August 9 and closes August 21.

Dr. Robert J. Wade is Camp Director and supervises a carefully selected staff of college-trained counsellors in a program designed to give handicapped boys and girls 12 days of enjoyable living and learning in an outdoor atmosphere.

The camp is equipped with all-weather cabins, a large dining hall, craft shelter, temperature-controlled swimming pool and out-post facilities, and there is a 110-acre lake for fishing, boating, and water sports. A physician and a registered nurse are on duty 24 hours a day and five student nurses from the Indiana University School of Nursing are on hand during each session to supplement the medical staff. Nondenominational religious services are held at the camp, but if the parents prefer, arrangements can be made to take children to nearby churches.

For children whose parents cannot afford the expense of their camping, campships are provided through interested individuals and groups connected with the Association.

The 170 physically handicapped campers enrolled in the camp this past year came from 57 Indiana counties. Thirty boys and 23 girls were in wheelchairs.

For information about Camp Riley or ideas to adapt their camp life to one for children the age of yours, write to the

Riley Memorial Association, 129 East Market Street, Indianapolis, Indiana.

An eating utensil for patients who require a mechanical apparatus to perform arm and hand functions has been developed by two employees at Highland View Hospital, Warrensville Township, Ohio, according to an article in the *Journal of the American Hospital Association.* The utensil was constructed by bending the handle of a swivel spork (a combined spoon and fork with wide tines) at a 90-degree angle one inch from the swivel. A one-inch metal plug was then screwed into the spork handle and a standard bicycle handgrip was attached to this plug. Because the compressibility of the handle allows a patient to obtain a more secure grip on the utensil, he is able to pick up the utensil independently, feed himself, and set the utensil down again. Previously, someone else was required to position a utensil in the patient's hand and remain with him throughout the meal to reposition the utensil if it was set down or dropped.

Barbara Fortune and Harry Fritz, the two Highland View Hospital employees who designed the utensil, were awarded a first prize for their idea in The Greater Cleveland Hospital Association Achievements Contest. It strikes us that you could easily get the help of a prosthetic appliance store in making something similar for the use of your growing handicapped child who may not be able to handle regular eating utensils.

According to an article by Joan Osterhoudt in the *Newark (N.J.) Star-Ledger* of June 13, 1971, of all parts of the body, the hand is the most frequently used and most prone to accidents. And Newark's St. Michael's Medical Center has a new clinic that specializes in the problems of the hand. Dr. Francesco Crocenzi, hand specialist there, considers the hand the direct appendage of the brain, and the clinic was formed in April, 1971, not only to help restore hands broken and mutilated by accidents and burns, but also those crippled by disease or congenital deformity.

The clinic works in close cooperation with the hospital's physical therapy, orthopedic, and plastic surgery departments, and Dr. Crocenzi said it is imperative that the patient be seen as soon after injury as possible, if maximum function is to be restored.

For every 1,000 births, six children are born with hand deformities. This may mean more than five fingers on one

hand, fingers that are fused together and must be separated by surgery, or "lobster claw" hands, where fingers never really develop. Of course, neurological and muscular diseases may also cripple the hands, and the construction of artificial joints will sometimes form part of the therapy, as well.

Most large medical hospital complexes are now able to handle problems of the hands, just as in Newark, and if not able to in your area, will be able to locate the nearest hand clinic to where you live.

Cystic Fibrosis:

While there is no cure for cystic fibrosis, new forms of intensive therapy, begun early, can now control the disease, prevent irreversible damage to the lungs and make it possible for most patients to grow into adulthood and lead relatively normal, happy and productive lives.

Of course, treatment must be individualized. Cystic fibrosis varies greatly in degree of involvement and severity from patient to patient and within each patient. The complications against which therapy is principally directed are bronchial obstruction, pulmonary infection, and pancreatic deficiency.

Especially important in the treatment of this genetic disease is the role of the physician. He not only must maintain close supervision and direct frequent check-ups over a very long period of time, but also should establish a particularly close relationship with the child and the child's family. Both children and their parents need constant encouragement and support as well as explanations and careful instructions in home care programs. Cystic fibrosis is a disease in which parent counselling plays an unusually significant part.

Production of abnormal viscid secretions (bronchial obstruction) in the lungs of patients with cystic fibrosis is a continuing process and must be combatted on a continuing basis. It results in a constant accumulation and stagnation of the secretions, dust, and bacteria, causing obstruction and inviting infection, which in turn increases the obstruction. Therefore the primary aim of therapy is to liquefy these secretions and to improve and maintain pulmonary drainage. Many experts advocate prophylactic treatment also, even when symptoms are minimal. This helps to prevent infection and the development of emphysema, atelectasis, and fibrosis.

According to the medical information booklet published

by the National Cystic Fibrosis Research Foundation, 202 East 44th Street, New York, New York 10017, (212) 687-0230, methods include aerosol treatments, nebulization of medications, mist tents, home humidifiers, postural drainage, breathing exercises, and physical activity.

Aerosol therapy—intermittent inhalation of particulate water and medications via nebulizers is effective in wetting and thinning the mucuous secretions in the lungs. This aerosol method also makes it possible to deposit directly on the mucous membranes of the tracheobronchial tree such agents as decongestants, bronchodilators, mucolytic agents, and antibiotics. It is often prescribed three to four times daily as both a therapeutic and prophylactic measure. Coughing and expectoration are encouraged before and during aerosol treatments.

Mist tent therapy—the use of a mist tent at night and during naps is of considerable benefit to cystic fibrosis patients with pulmonary symptoms. Many physicians believe it to be the most effective way to improve and maintain optimal pulmonary hygiene and function even in patients with minimal evidence of airway obstruction.

Tepid mist therapy—patients with acute or advanced chronic pulmonary disease benefit from mask aerosol treatments with a unit which delivers a 98° mist. It provides maximal wetting of lung secretions.

Home humidifiers are of value especially during periods of low humidity in the summer and when the heating system is in use during the winter.

Intermittent positive pressure breathing therapy should be used only in special cases and under the supervision of an experienced physician.

Physical therapy—postural drainage. Used in conjunction with aerosol therapy, postural drainage is one of the most important forms of therapy for patients with cystic fibrosis. Careful positioning is important to achieve efficient drainage of affected lobes. The physical therapist uses chest-clapping and vibration procedures for each position while the child is encouraged to cough up as much sputum as possible. This therapy is carried out twice daily whether the cough is productive or not. To be effective, postural drainage must be taught to the parents (while the child is young) so that it can be carried out at home, and to the child later when old enough to do it alone or with the help of parents. Special

training for physical therapists in these techniques is essential.

Physical therapy—breathing exercises. Children with breathing difficulties get in the habit of using accessory muscles for respiration and so tend to develop rounded shoulders and pouter-pigeon breast. The purposes of breathing exercises, which can be taught by the physical therapist, are to improve efficiency of respiration and ventilation and to improve posture.

Details on recommended nebulization equipment and on methods of inhalation therapy can be obtained by physicians directly from the National Cystic Fibrosis Research Foundation.

Physical activity—Running, swimming (no diving), and active play can often do more to help raise sputum than other forms of therapy. Therefore, physical activity should be encouraged and confinement to bed avoided, if possible.

Environment—Humidity is important and should be maintained above 50 per cent where possible. Home humidifiers are recommended, as is air-conditioning, in hot weather. A mild climate may be helpful and is preferred over a cold damp climate where respiratory infections are frequent in the general population. But the results of a change in climate are not sufficiently striking to justify moving to a different area.

Precaution—At no time should medications be administered that suppress the cough reflex.

The treatment of pulmonary infection is best left to the individual physician. The best method to prevent infection is therapy aimed at keeping the lungs clear and because respiratory infections often develop and progress insidiously, early detection and vigorous treatment suggested by the physician are of paramount importance, which is why frequent clinical evaluation is essential. For many patients, even monthly checkups may be necessary.

The various organisms responsible for infection in cystic fibrosis are all noted for their tendency to cause low grade chronic infection and to become resistant to antibiotics. However, your physician will know the best kind of antibiotics to help your child during such an infection, and he will be the one to decide upon the method of their application.

Pancreatic enzyme deficiency is treated by replacement therapy and diet. Extracts of animal pancreas (pancreatin) are given with each meal. Both the preparation used and the

dosage must be determined for the individual patient by clinical trial. This treatment results generally in improved digestion and absorption of fat and protein, reduction in number and bulk of stools, improved weight gain, and decrease in excessive appetite. It also helps to prevent or control prolapse of the rectum.

The diet does not need to be rigidly restricted but should be high in protein and low in fatty foods. It may have to be adjusted to the tolerance of the individual patient. Water-miscible vitamins, especially vitamins A and D should be given in twice the usual dosage at all ages. There is still some question as to the necessity of adding vitamin E to the diet. Although symptoms of vitamin E deficiency have not been shown in man, the serum of patients with cystic fibrosis is low in vitamin E. It can be given in water soluble form at a dosage of 1-5 units or milligrams per kilogram daily.

Treatment of the sweat gland defect—All children with cystic fibrosis lose excessive amounts of both sodium and chloride in their sweat. They need to be protected from acute salt loss during hot weather, febrile periods, and periods of strenuous physical exertion. They should be given extra salt with their food throughout the year and may require salt tablets in hot weather.

Surgical complications—Surgery usually is necessary to correct meconium ileus. Between five and ten per cent of patients with cystic fibrosis have this condition—intestinal obstruction—at birth. Putty-like, black meconium obstructs the lumen of the small intestine. Unless corrected by surgery, the condition proves fatal. Patients who survive will develop the symptoms of cystic fibrosis and can expect the same course and outlook as those who have not had meconium ileus.

Surgery may also be necessary in certain other conditions resulting from cystic fibrosis, such as: (1) surgical shunt procedures for cirrhosis with portal hypertension and esophageal varices; (2) thoracic surgery to resect localized areas of extensive bronchiectasis or abscess; (3) surgical intervention when pneumothorax and emphysema occur, and (4) ear-nose-throat surgery in the form of adenoidectomies, resection of nasal polps, and sinus drainage.

When any surgical procedure is performed on patients with pulmonary involvement, the selection of pre-anesthetic and anesthetic agents must be made with great care. Every effort has to be made to maintain a clear airway. Morphine,

codeine, and other drugs that suppress the cough reflex should never be used.

Treatment of cardiac involvement—Symptoms and signs of right heart failure may appear acutely during severe flare-ups of the pulmonary disease or may be persistent with far-advanced pulmonary disease. Of primary concern is intensive treatment of the underlying pulmonary disease, employing all the measures for pulmonary hygiene discussed earlier, such as postural drainage, mist tent therapy, breathing exercises, and other such measures. The use of oxygen (with great caution), digitalis, mercurial diuretics, and rest may also provide the time needed to bring the pulmonary disease under control. Usually, when this is accomplished, treatment of the cardiac symptoms may be discontinued.

Many cystic fibrosis patients show a grayish discoloration of the teeth. It is believed that this is due to prolonged use of broad-spectrum antibiotics, especially the tetracyclines.

In general, many variations in the severity of these signs and symptoms of cystic fibrosis are seen. Many children show severe disease from the first weeks of life. Others may not have symptoms until 18 months or two years of age, or even later. In some cases there is marked pulmonary disease without any symptoms of pancreatic or other digestive involvement. On the other hand, the gastrointestinal manifestations may predominate for months or years before any signs of chronic pulmonary involvement appear.

Cystic fibrosis is *not* contagious. Children with it may attend school and engage in normal activities insofar as their condition permits. They should be protected as much as possible from exposure to infection and should be given all the routine inoculations, including smallpox, polio, diphtheria, tetanus, and pertussis (whooping cough). Measles and other viruses which affect the pulmonary system can produce extremely serious involvement in patients with cystic fibrosis. Measles vaccine should be given and measles itself should be modified with gamma globulin. Influenza vaccine should be administered during epidemic periods.

Advances in treatment for cystic fibrosis in recent years have greatly improved the prognosis for both morbidity and mortality. Because of the variations in the severity of the disease, the outlook also varies from patient to patient, and is unpredictable for the individual patient, especially in infancy. Optimistically, with intensive prophylactic care, most pa-

tients with cystic fibrosis who are diagnosed before irreversible damage has occurred in their lungs will live until adulthood.

This outlook will become true for more and more patients as awareness of the disease and its symptoms spreads and as more patients are detected in the early stages. Because of its chronic and exceedingly complex nature, cystic fibrosis requires the kind of comprehensive treatment and total care approach that is best provided by a team of experienced workers familiar with the disease. Such teams are available at the Cystic Fibrosis Care, Research and Teaching Centers listed in your local telephone directory, or your nearest one secured by writing to the national office listed earlier in this section.

These centers are very important to families with a cystic-fibrosis child, not only because of the important role that is played by nurses, physical therapists, and medical social workers in helping to provide care and support. The burden emotionally, socially, and financially on families can be enormous. The cost of drugs, diet supplements, and equipment alone often exceeds $1000 per year, year after year, and is frequently more than the average family can bear. The need for constant supervision and for daily home therapy programs, with the danger of life-threatening flare-ups always present, can bring despair. Through their professional services and by offering understanding and encouragement, plus information and guidance to obtain needed assistance from various community resources, medical social workers can provide aid of inestimable value.

Incorporated in 1955 and consisting then of a few scattered groups of parents of children with cystic fibrosis, the National Cystic Fibrosis Research Foundation is now a rapidly growing national voluntary health organization with 10 chapters in 48 states. It supports programs of research, care, education, and training for scientists, physicians, and personnel from the allied health professions and maintains chapter service programs for patients.

These programs are financed primarily through funds raised by its chapters. Chapters retain 25 per cent of net funds to finance local service programs; 75 per cent is used to finance the Foundation's overall national programs, including grants for cystic fibrosis centers, research, research fellowships, and professional education.

Until 1960, the Foundation's funds were used almost exclusively for research grants, but in 1958 funds for such research became available through the U.S. Public Health Service-National Institutes of Health. A study of the total cystic fibrosis problem exposed other major needs and gap areas not filled by any other organized effort, including a nationwide network of care, research, and teaching centers on cystic fibrosis; local diagnostic and treatment consultation clinics; educational services and materials; chapter patient-service programs to provide assistance with equipment, drugs at reduced prices, special services through clinics and other institutions, and chapter programs to guide parents to community resources for home nursing, treatment facilities, teachers for home-bound patients, assistance available from private and public agencies, plus a chapter information program providing educational services and materials designed to awaken public interest in cystic fibrosis.

Most other volunteer health groups that have been established for some time provide similar services, and can be located through your local telephone directory, your doctor, or your County Medical Society.

Mentally-Retarded, Emotionally-Disturbed and Brain-Damaged Children:

It is known to science that at least three per cent of all American children (well over one million in number) are mentally retarded, and that 2,100 of them are born each week who will be mentally retarded. This means that mental retardation is six times as frequent as epilepsy, ten times as frequent as diabetes and 20 times as frequent as muscular dystrophy.

Unfortunately, many parents prefer not to admit to themselves at first that their child is mentally backward, until they learn through a teacher that he is not keeping up mentally with his peers, other children his age.

Psychological testing provides the final, positive evidence that a child is retarded. It presents parents with facts that cannot be ignored or reasoned away.

A few years ago, Dr. Augusta Goldin wrote a fascinating article in the *Sun Science News* (we regret not having the name of the city in which this paper is published), stating that it is fortunate that the vast majority—85 per cent—of

mentally retarded children turn out to be only mildly retarded.

"They are classed as 'morons', an unfortunate word with connotations as heartbreaking as the knowledge that a child is subnormal. However, these children have an I.Q. ranging from 50 to 70 and are capable of learning limited academic and vocational skills. They can earn a living and take care of themselves.

"Children with I.Q.'s ranging from 20 to 49 are referred to as 'imbeciles' and represent 11.5 per cent of the retarded group. The remaining 3.5 per cent of these children possess I.Q's of 0 to 19 and are severely subnormal. Most of this latter category, classed as 'idiots,' together with the imbeciles are institutionalized at birth. The remaining ones are committed some years after their parents die or can no longer take care of them.

"Mental age and intelligence quotient," continued Dr. Goldin, "are determined by administering a battery of tests. If a child is a poor reader, he or she is tested verbally. Testers use standardized pictures, drawings, and objects to check out language abilities, reasoning abilities, and understanding of spatial relationships.

"A child's score represents his mental age in months. If, for example, he scores 96, his mental age would be 96 months. Intelligence quotient is computed by dividing this mental age by chronological age.

"If a child is exactly eight years old, or 96 months, his I.Q. is a normal 1.00 or 100. If an eight-year-old scores 80, this mental age divided by 96 comes to an I.Q. of 83 or 83. An I.Q. of 83 places a child in the normal, but dull, slow-learning range.

"If a boy or girl has an I.Q. of 75 or below, he or she would be placed in a special class for children with retarded mental development. Here, at a slowed-down pace, the child is provided with individual help . . ."

According to Adam P. Matheny, Jr., Ph. D. and Joel Vernick, National Institutes of Health, Bethesda, Maryland, in an article in the *Journal of Pediatrics*, 74:953-959, 1969, condensed in *Modern Medicine*, in the January 26, 1970, issue, parents of mentally-retarded children need specific, clearly transmitted, honest information about the child, implications for his future, and knowledge of what concrete steps they can take to effectively deal with his problems.

"In many instances," the article stated, "refusal on the part of the parents to accept limitations on their retarded child's life results from nonemotional constraints—their level of education, lack of knowledge about retarded children, existing expectations based on normal children, and inability to translate the child's present abilities into future capabilities. . . ."

Brain-injured children, however, are *not* retarded children. "They have I.Q. marks comparable to, and often higher than those most would call normal children. They do not suffer from physical abnormalities and could not be picked out of a crowd of children," according to an article in the *Newark Star-Ledger*, January 5, 1969, by Robert J. Braun, that paper's Education Writer.

"But they do not—they cannot—learn properly," he continued. They have a very short attention span. Perhaps they cannot sit still for any long period of time. They may be able to get along with other children or their teachers," he quoted James Allegra, a Hohokus, New Jersey, special education teacher. From the best information available, Allegra said, "something" happens to a child's perception process. Although he may see printed letters and words, the transmission of what these printed symbols mean never makes it to the child's brain. What that something is, how that scrambling of meanings works, is still very largely a mystery.

"That's why a brain-injured child looks no different from what you would call a normal child," Mr. Allegra was quoted. "But put that youngster in front of a book and the difference becomes immediately obvious. . . ."

Mr. Allegra feels that parents can do a great deal to improve their children's learning capacity. The first thing parents must learn if the child is brain-injured is that he does have a problem. The possible development of a neurologically impaired child cannot be predicted, the teacher said.

Worse yet, the parents may be tempted to distrust the public school educational classes and place the child in an institute. "They just do not understand," said the teacher, himself the father of a young child, "that parental love is perhaps the most important thing this child can have. An institution means visits on weekends only—maybe less than that. It should be taken only as a last step, the very last step."

Once parents accept the problem for what it is, they must

provide structured activities, meaningful schedules of household chores, and in some cases, specialized tutoring for their children. "The child finds safety, comfort, security, and reassurance from the reliability of enforced organization, even though he may balk at times," the special education teacher counselled through Mr. Braun's article.

He urged parents to treat their neurologically impaired child exactly the same way they would treat a normal child of the same age. These children should be allowed to participate in social gatherings—although these must be carefully scrutinized. These children, he explained, have had so many discouragements in their lives, they should not be put into a position which would cause more failure.

The young special education teacher also tries to break down the barriers between the normal children at the Hohokus public school and the students in the special class for the brain-injured. "Let's face it, young children are cruel. They will do anything to youngsters—from calling them names to freezing them out of common play," he said in the article.

He has tried to develop what he calls "sharp blows to the stigma of neurological impairment,"—for example, he has arranged for the special students to learn how to type. "Now these kids can type where the children in regular classes cannot. In a way, they are a step above the rest. These techniques help to stop the teasing—and they also serve to build confidence in the special-education child."

Writer Robert Braun ended his article by asking: "What is in the future for these children, children who seem to be neither mentally retarded nor physically impaired?"

"It has to be promising," Allegra answered. "All you have to do is look at their condition ten years ago. They were completely forgotten people."

Two organizations which are specifically concerned with minimal brain dysfunction in children are: The California Association for Neurologically Handicapped Children, P. O. Box 604, Main Office, Los Angeles, California 90053, and The Association for Children with Learning Disabilities, 2200 Brownsville Road, Pittsburgh, Pennsylvania 15210.

If your child is not called "brain-damaged" but the other phrase, "minimal brain dysfunction," is used, be encouraged by the fact that Dr. Miriam W. Carmichael, neurologist of Richmond, Virginia, in an issue of *Medical Tribune* defined this as usually referring "to children of about average intelli-

gence who evidence learning or behavioral difficulties thought to be related to abnormal function of the central nervous system." As mentioned in the proceding section of this book, examples of symptoms of minimal brain dysfunction or minimal brain damage include inattention, hyperactivity and other behavioral abnormalities, clumsiness, speech disorders, perceptual handicaps, reading problems, emotional disturbances, and others.

One of which we hear quite frequently these days is hyperactivity or hyperkinetic behavior, and this also was described in detail in the preceding section. However, hyperactivity is actually a general term that means different things to different people. According to a booklet published by Smith Kline & French Laboratories of Philadelphia, Pennsylvania, pharmaceutical manufacturers, when a physician uses the term "hyperactivity" he is describing behavior relative to other children of the same age. To the parent, the term is used to describe behavior that often seems to demand excessive discipline in the home. To the teacher, a hyperactive child is one that creates problems in the classroom. In all cases, hyperactivity refers to behavior that is different from the behavior of the average child of the same age.

What causes hyperactivity? A number of theories have been advanced, says the Smith Kline & French booklet. The explanation most widely accepted at present is that hyperactivity occurs when the control centers of the brain have not developed as rapidly as those centers that cause mechanical function. Intelligence is not impaired though emotional immaturity is common. One thing is certain, however. While brain damage may cause similar symptoms in some children, most hyperactive children do *not* have histories of brain damage. Neither does hyperactivity run in families. More boys than girls and more first-born than later children are hyperactive, according to available statistics.

The booklet continues its explanation by stating that hyperactivity is usually diagnosed and discovered when the child begins nursery school, somewhere between three and four years of age. In the majority of children who receive treatment, the hyperactivity diminishes gradually as the child matures and usually is no longer a problem by the time the child reaches adolescence.

Some early indications may appear in infancy, however. Head-bangers and crib-rockers may be hyperactive. Later on,

teeth-grinding and temper tantrums can be symptoms. There is nothing rare about hyperactivity in children, surprisingly enough. For example, in the public schools of New York City, it is estimated that about four per cent of the total enrollment (that's 44,000 children) have been diagnosed as hyperactive. And conservative estimates of the entire elementary school population of the United States indicates that over a million children are hyperactive.

There are several different kinds of drugs used today to help treat this kind of child. If your physician has prescribed one of these, it is very likely either a member of the tranquilizer group or one of the stimulating drugs. Some hyperactive children respond best to one type; others respond better to the other. Such drugs can make a hyperactive child's conduct more normal. Tranquilizers tend to slow him down and make him more manageable. In addition they help a child control his emotions. Stimulating drugs, such as the amphetamines, are currently the most frequently prescribed agents for the hyperactive child—both as treatment and as an aid in diagnosis. As mentioned before, it is believed that in the hyperactive child the control centers of the brain have not developed as rapidly as the motor centers. So, in some children, by stimulating the control centers, these drugs enable the child to have more control over his behavior and therefore to function more normally. As a rule, the hyperactive child will respond to drug therapy within a few days.

If no improvement is noted in two or three days, call your physician for advice. Usually, all that's needed is an adjustment in dosage of the medication. But *do not* attempt to change the dosage yourself. Stimulant drugs have been used successfully to treat hyperactive children for over 30 years. This experience has shown no tendency toward habituation or addiction in children. Occasionally, a few children taking stimulant drugs may develop a pale, anxious expression. However, the most common side effects are insomnia and reduced appetite. These are not usually a problem if the drug is given just before breakfast and before lunch. But, except for individual sensitivity, which your doctor will probably mention, experience has shown that hyperactive children usually tolerate stimulating drugs better than adults.

If you notice a side effect—or no effect—notify your doctor immediately. Again, a change in dosage may be needed or even a change in medication. But never, under any circum-

stances, should you attempt to decide to make such changes yourself. Only your doctor can make this decision for your child.

There are many ways in which you can help your hyperactive child to slow down. Your goal is to eliminate as many frustrations as possible, says the Smith Kline & French booklet, by helping him to re-channel his energies constructively. Begin by asking yourself a question. Are you expecting too much of your child? It's natural for parents to want their children to excel. But perhaps your sights are prematurely high. Don't push. Relax. Try not to be tense. A child is quick to sense impatience and your attitude or actions may aggravate his behavior.

Anger is a normal and frequent byproduct of hyperactivity—both in your child and in yourself. So be prepared! Expect your child to be angry when frustrated and remember that his frustration level is abnormally low. Let this foreknowledge help you to handle his outbursts sympathetically while keeping your own anger under control.

The hyperactive child is more comfortable on a fixed routine so that he can expect certain things to happen at a certain time—mealtimes, naptime, playtime, TV-time, storytime, bedtime, and so forth. He will also need forewarning of even the expected events. If it's playtime, say "Now we will play with this." And play with him, giving him your attention. If your attention wanders, he'll wander. Though he may not show it, or be as responsive as you might like, he needs a little extra love and extra attention. Order and regulate your home life so that you can give him the attention he needs.

Obviously, no family's activities can or should be inflexible. If, for example, you have agreed to watch television with him at a specific time and then find you must go to a PTA meeting, explain to him why you can't and offer him an extra bit of time the next day.

Temper tantrums of a violent nature should be handled in the same way as with any other child. At their peak, restrain him quietly but firmly and prevent him from doing damage or harming himself. When his anger subsides, point out what he has done and offer comfort and soothing attention. Frequently you will find that he is ashamed for having lost control and needs your comfort and reassurance.

How can his older brothers and or sisters help him? His

problem should be explained to them so that they will understand and be able to help in minimizing tension and frustration. However, very young children quite naturally may poke fun at him, even scoff and sneer.

What about grandparents, close relatives, and friends? Their patience and understanding should also be encouraged. Grandparents, however, tend to be difficult, and their attention may stimulate the child to greater hyperactivity. If this is the case, keep calm and if necessary enlist their cooperation in limiting the length and frequency of their visits.

The play activities that are best for the hyperactive child depend on his age and his abilities. What is important in any activity is that what he does must hold his attention and usually this will require the help of another person. To give the hyperactive child paper and fingerpaints and expect him to play at this alone is unreasonable. Make a game of it. Do it with him. Teach his older brother or sister how to help hold his attention. *Supervision means that you care about him.* And that he needs to know. Choose playthings and activities that are right for his age, that are noncompetitive and that do not require concentration or skills that are beyond him. Build his confidence by giving ample time—and priase—for things he seems to do particularly well. Remember, he becomes bored very easily.

Games that lend themselves to numerous variations are particularly desirable, such as ball games, for one. The variations possible from infancy on are almost endless. Check any good child-development book for what game at what age. Or simple jig-saw puzzles. The hyperactive child usually has good coordination and learns physical activities faster than other children. Swimming and other noncompetitive sports provide an excellent way of letting him throw off a lot of steam. You must not get him into competitive games, even with children his own age, until his hyperactivity levels off, since he is in enough competition with himself. Games involving groups of children or waiting his turn may overstimulate him or increase the chance of frustration, compared to those that can be played with fewer children or enjoyed individually. Later, when he's improved and has gained more self-control, he may gradually be encouraged to join his peers in competition.

With the cooperation of your doctor, your child's teacher, and your family, the chances for the hyperactive child's fu-

ture is a good one. Close yet relaxed supervision on your part, working together with your physician, will help channel and direct your hyperactive child's resources and to provide suitable outlets for his excess energy until he is able to assume responsibility for his actions. Usually, with the proper care and treatment, hyperactivity ceases to be a problem by adolescence.

The Bureau of Education for the Handicapped (this can be mental or physical handicaps) has started a new service in behalf of children needing special education. They've computerized the name, address, and nature of the services of every educational project in the United States which offers programs for handicapped children. This information is available without charge to parents, guardians, teachers, and other interested persons. Send your request to "Closer Look," Box 1492, Washington, D. C. Also, the National Association for Mental Health, Inc., 10 Columbus Circle, New York; New York 10019 has an affiliate in each state, 850 local chapters, and sponsors a number of programs for mentally ill children. Last, The Children's Bureau, U.S. Dept. of Health, Education and Welfare, Washington, D.C. 20201, ATT: Information Dir., can give you direction for anything you need to know about mental health problems.

As far as the educationally-handicapped child is concerned, the July 26, 1971, issue of the *Journal of the American Medical Association* had an excitingly helpful letter from Elaine Bierbauer, RN of Santa Ana, California, calling attention to the fact that it has been estimated that approximately five million of America's school children suffer from some degree of learning disability due to minimal brain dysfunction, and that it is not enough for the physician to diagnose and treat the disorder, but he must be able to advise and counsel parents in the home management and education of these children.

The letter advised that 42 of the 50 states have some kind of parent-sponsored organization which provides information and help to parents of these children, such as the California Association for Neurologically Handicapped Children which sponsors parent-education programs, provides literature and films, and promotes legislation for programs of special education. Nurse Bierbauer also stated that the Association for Children with Learning Disabilities is a national organization similar in nature, has 200 chapters throughout the country,

and that information can be obtained from Mrs. Robert M. Tilottson, Secretary, 2200 Brownsville Road, Pittsburgh, 15210. This national organization, as well as the California one mentioned, publishes informative newsletters for their members.

The booklet, "Learning Disabilities Due to Minimal Brain Dysfunction," can be obtained from the Supt. of Documents, U.S. Government Printing Office, Washington, D.C. 20402 at a cost of $15 per 100 copies, or 20 cents for individual copies, and a reading list of books is available on this disorder.

Nurse Bierbauer's letter concluded: "Providing parents with an adequate understanding of the nature and treatment of this disorder along with information about the availability of community resources makes it possible for the physician to provide total care for these patients."

Asthma:

The child born with asthma sometimes seems to live in an endless cycle of limited activity, doctors, hospitals, oxygen tents, and powerful, life-sustaining drugs. Many are never even permitted to walk to school and most of them are put on a daily regimen of steroid medications which, while helping to keep them alive, robs them of their natural growth.

One organization has been around for years trying to demonstrate that under the right medical and institutional program, asthmatic children can be nearly as healthy and normal as anyone else—if only parents will understand that there is a limit to the help that can be given such children at home.

Doctors at the Children's Asthma Research Institute and Hospital (CARIH), 3401 West 19th Avenue, Denver, Colorado 80204, feel that a child with asthma relaxes better when he knows he will find everything needed for a severe asthma attack that might hit him. Not being afraid or anxious is one of the most helpful parts of asthma therapy for a child. At the institute, as their attacks decrease, they begin to believe that perhaps some day their terrible illness will become just an occasional nuisance or merely a bad memory.

Parents, particularly mothers, cannot help panicking whenever their child has an asthma attack, loses control of his breathing, and has to be rushed to a hospital. Such fear is contagious, and when everyone around the child is anxious

and acting as though he's about to die, or else lets him remain undisciplined, they bring him additional problems in his life.

Avoiding panic and anxiety in the child is very difficult when he is wheezing heavily. At CARIH, he learns to breathe in a relaxed manner during attacks, and it is easier when he sees the example of other children, like himself, going through the same procedures—and attacks—that he does.

At the Denver Institute, patients usually progress to the point where they can finally indulge in winter sports, mountain climbing of sorts, and all kinds of spectator activities. They learn that they can make an asthma attack more or less severe for themselves, become hospitalized, or they can learn to be calm and prevent each new attack from becoming as severe as the last one.

Children learn that if they get three bronchodilating inhalations (called "nebs" because a nebulizer spray gun is used) in any one day, they are automatically hospitalized, and that two nebs means restriction to the grounds. When there is an activity scheduled in which they want to participate, they try to remain calm, because they don't want to take nebs and be grounded.

CARIH staff members feel that regular discipline is just as important to the asthmatic children as medical care. They also believe that having a chronic illness should not change the rules of behavior of a child, or sharing with other children.

It costs about $7,000 a year to care for a child at CARIH, but parents do not have to pay anything, as the Institute is supported by private donations garnered by a nationwide network of 140 auxiliary chapters. Of course, those parents who can afford it may make whatever financial contributions they wish. The Institute is able to care for approximately 100 children, plus staff, all of whom live in six cottages spaced around the 32-bed hospital and research facilities on the 17½-acre site in West Denver.

If you wish further information on asthma, write to the National Institute of Allergy and Infectious Diseases, Office of Information, Bethesda, Maryland 20014. As you can see from their title, they also cover other problems besides asthma, so in the same letter, why not ask for information on allergies, as well?

Epilepsy:

Shame and guilt are two very harmful parts of caring for the epileptic child and do *not* belong in his regimen. A child can sense distaste, even when not expressed verbally, and epileptic children require love, affection, caresses, and reassurance just as much as any other sick child, possibly more, because they frequently are not even aware of their seizures, and would not understand the reasons for the faint hints of shame from family members or others.

According to the national organization, The Epilepsy Foundation of America, 733 15th Street, N.W., Washington, D.C. 20005, which can direct you to its local and state chapters in your area, many epileptics can lead normal lives, if their families do not feel shame or guilt for their illness.

Persons who witness a child (or an adult) having an epileptic seizure should observe the following advice:

(1) Don't panic; keep calm.

(2) Help break the patient's fall.

(3) Move away objects on which he can hurt himself, or else slide him away from them, if easier.

(4) Don't force something into the mouth of an epileptic having a seizure in the belief that you are helping to keep his tongue from choking him. Instead, either put a soft handkerchief in the mouth, not too far back, or else gently turn the head of the patient to one side, so he will not choke on his saliva.

(5) Do not talk to or bother the patient. Usually, he will come out of the attack within a few minutes and may not even realize he has had one.

(6) Never force water or medication down the throat of an epileptic while he is having a seizure or recovering from one.

(7) Do not take him to a hospital unless the seizure lasts as long as a half-hour and he does not seem liable to gain consciousness normally. Otherwise, you'll find a wasted ambulance trip, because the patient will be his normal self again when the ambulance arrives.

The spring, 1971, issue of *The National Spokesman*, the publication of the Epilepsy Foundation of America, put its spotlight on the Epilepsy Center of Michigan (ECM) as an

indication that strong and viable epilepsy state-groups can be developed.

"In 1970, ECM provided direct services to nearly 5,000 persons in 82 counties throughout the state," said the article. "These services included diagnostic studies, medical consultations, information and referral activities, and counseling. . . .

"The ECM patient care program begins with evaluation services which may include both diagnosis and initial seizure management. The evaluation consists of neurological examinations, interviews with social workers, psychological testing and interview, psychiatric examination, EEGs, skull X-rays, and other laboratory tests, as indicated.

"Following the evaluation, the staff diagnostic team makes its recommendations at a conference to which the patient's personal physician and professionals of other agencies working with the patient are invited. An 'interpretation interview' is then held to review the staff recommendations with the patient and his family.

"While there is generally a fee for diagnostic evaluations, fees are based on the ability to pay, and no Michigan resident is denied services because of inability to pay.

"There is no charge for other patient services. These include a statewide counseling program designed to assist patients and their families to make a positive adjustment to epilepsy. . . ."

There are many other services offered by not only the Michigan Epilepsy Center, but those in other states, and most of them have clinics in the larger cities of their states to help treat such patients.

Epilepsy is a long-term disorder and the continuous use of medicine over many years imposes a real financial burden on almost any family. In 1968 the National Epilepsy League Service, 203 North Wabash Avenue, Chicago, Illinois 60601 began a pharmacy program that has saved many victims of that disease (or their families) at least 25 per cent on their pharmaceutical needs. It is not a business (no profits accrue to any individual) and it is not a charity (members share the cost and benefits equally). The highest ethical and professional standards have been set for the program, which works in the following manner:

If someone in your family has epilepsy, prescriptions

mailed to the National Epilepsy League Service, 203 North Wabash Avenue, Chicago, Illinois 60601 will be handled if you observe the following instructions:

(1) The prescription must be made out and signed by your own physician. (Do not ask for the one your pharmacist has on file, because he is required by law to hold on to it. The League does not provide medical diagnosis or treatment or advise the medicine prescribed.)

(2) Orders will be filled for any kind of medication prescribed in the treatment or management of epilepsy, including anticonvulsants, tranquilizers, headache remedies, and antibiotics. Medication unrelated to epilepsy must be bought from your local pharmacy.

(3) The National Epilepsy League Service stocks only fresh medicine of the highest quality manufactured by ethical pharmaceutical firms. No drug products are made specifically for NELS.

(4) Medicines are mailed promptly and in plain wrapper. All incoming and outgoing mail and checks or money orders must be marked simply "NEL Services."

(5) Always order at least two weeks before your child's supply of medicine runs out. If a new prescription is required before you reorder you will be notified, and your doctor must state on the prescription whether it can be refilled.

(6) Only League members with the disorder of epilepsy are entitled to participate. Of course, parents or guardians are permitted to order for an epileptic child. Membership dues for this special services are $1.00 per year.

(7) Send no money in advance. The League will send you a bill with the medicine. You pay 25 per cent less than the usual price (plus the $1.00 for membership if it is your first order for the year). Everyone is expected to pay the actual billing cost of his medical order promptly, no later than within 30 days.

By joining this cooperative plan, prices for medications for epileptics are cut for everyone, including those usually unable to afford the effective but expensive anticonvulsants. The Board of Directors of the National Epilepsy League Service has stated that all epileptic Americans are invited to take advantage of this reduced-cost pharmaceutical service.

Most epileptics will have fewer seizures if they are allowed to lead normal lives. That is why your child should be allowed to have friends, go to nursery school or kindergarten, and should be treated as a normal person. If it appears necessary to make any explanations for the lack of alarm by you over your child's condition to the parents of his young friends, or to your neighbors, secure material from your local epilepsy group chapter, or ask your child's doctor for a letter you can circulate from him, to explain the harmless nature of your child's condition.

Education, of course, should begin with you and your family. You should all become completely knowledgeable as to everything about epilepsy and each of you should participate in some way in the volunteer work of the epilepsy groups.

Today, epilepsy is seldom a fatal disease. It is a problem, of course, but not half so bad a problem as many of the other birth defects. When you permit it to assume its rightful place in your life and that of your family, epilepsy can prove to be just something else you are able to cope with as part of the program of your daily life.

Hearing and Speech Problems:

Since many birth defects result in hearing problems that range from slight to encompassing, it is important to remember that the first order of life with *any* child is to learn how to communicate with him.

With a deaf child there are two ways—both sign language and lip reading. However, the best lip reader in the world can only understand about 40 per cent of what is being spoken, as usually those doing the talking with their lips are not aware a lip-reader is around, and they do not slow down to accommodate him. That is why sign language is probably the most important method of quick communication between your deaf child and others.

Lip reading, of course, has another function besides attempt at communication with hearing people. It is a way of preventing the deaf child from feeling self-conscious in public due to his handicap. When lip reading alone is taught to a deaf child, it is called the straight oral method.

The first five years of his life are those in which you and your child must concentrate on his communicating with others. Children who learn sign language first and lip reading

second are able to speak better themselves, because they are not under the strain of partly comprehending, as in lip reading. But for some unknown reason, many hearing people feel a sense of exposure and shame when they must use sign language in public with someone who is deaf. There should be no sense of shame attached to communicating with a handicapped child, and the first examples of this should be the family members, especially the parents. When you are not constrained or embarrassed about what others think when you use sign language in public with your child, then your child will feel easier about his handicap and will probably do better, too.

According to various schools for the deaf, one out of every 1,000 persons in the United States is deaf, and most of those from birth or early childhood. It is advised that to communicate at night with a deaf child who is in bed and in the dark, you first take his hand, or else turn on a bright light, in an attempt to have him lip read or sign talk, should he be frightened.

The modern approach is to teach your deaf child from the time you become aware of his problem. Use a hanging mobile over the baby's crib, to encourage him to keep making sounds and thus learning to use his voice, even though he can hear nothing for himself. Consciously repeat gestures and words and actions for your deaf baby, again in spite of his not being able to hear you. He may try to imitate you, and he will react more normally to affection and hugs and games, when he knows you are reaching out to him, and not letting him lay or sit like an inactive doll.

In some cases of deafness, your doctor will send you to an otologist to see whether your child may be fitted with a hearing aid. The otologist will use an audiometer to measure your child's ability to respond to various depths and types of sounds and tones, thus telling whether the hearing loss is too great for help with a hearing aid, or whether one type or another will do him some good.

The otologist will also be able to tell you of local and state programs for your child—whether recreational or medical—and other ways in which his school and home life may be improved.

Since so much of learning to speak is hearing sounds, then associating and repeating them, teaching a deaf child to communicate who cannot use a hearing aid is even more of a

problem. But it can be done, even though the vocal tone is not as lively as with a child who can hear a little.

Nearly every city has a hearing and speech center of some kind, which can be found through your doctor or through your local health department or county medical society, and it is these places which can give parents and families help in working at home with their deaf child, to improve his speech and communication.

One good thing is that except in cases of multiple handicaps, such as from the rubella epidemics or from a particularly bad case of cerebral palsy, deaf children seldom have other serious handicaps. It is false for people to believe that because a child cannot hear well or at all, that he is automatically mentally retarded.

You must not postpone taking your deaf child to a nursery where there are other deaf children. Many hospitals are now equipped to test for deafness soon after birth, and as soon as you know of his problem, his only means of communicating with the outside world will be through the help you get for him.

A wonderful example of an excellent nursery school for deaf children was written about by Lois Lachstadter in the Dayton (Ohio) *News* of May 31, 1970. The school is a part of the Hearing and Speech Center of Dayton, and takes children as young as 13 months, although the center, itself, provides therapy for older children and adults, as well. Tuition at the nursery school is gauged according to the parent's ability to pay, but no one is ever turned away. The method of teaching is excellent:

"Seated on the floor with four deaf children under the age of two, Miss Karen Postma, speech therapist, thunders out, 'Shoe! Shoe! Give me the shoe!'

"A tiny doll's shoe is clutched by one girl. She hesitates a moment and an impatient classmate (age two) lip reads, understands the command, and spontaneously pulls off his own shoe and hands it to Miss Postma.

"This is followed by an exercise in association where one child grasps an apple, says the word 'apple' then runs to touch it to a picture of an apple mounted low on the wall.

"Another youngster puts her hand on Miss Postma's throat to feel the vibration as the young therapist says the word 'happy.' The girl squeals with understanding and repeats, 'Happy!'

"When the correct answers come, they're greeted with praise and acceptance from the young therapist, a graduate of Kent State whose interest in children dates back to age 13 when she started her own back-yard nursery for 14 neighborhood youngsters.

"The class ends with a warm, 'Want to hug good-bye?' as each child rushes to embrace Miss Postma. Minutes later four more children file into the room. The voice urges, emotion flows as the next class begins.

"Afternoons bring more classes as well as private therapy sessions at the center when Miss Postma works with one child at a time. After five, on her own, she visits some of the children at their homes for private tutoring.

" 'Repetition and consistency—they're so important. You have to keep repeating and repeating to teach these children,' she explains. 'Work! Work! Work! You keep working. Their little minds are so responsive. It's really amazing how quickly they can pick things up.'

"With these preschool tots the goal is clear—to bring them, by the time they are mentally four years old, to a level of communicating that will enable them to enroll at the Kennedy School for the Deaf. If they excel there, they may be permitted to enter a regular school with hearing children.

"Results come only after hours of work—not only in class and in private therapy sessions but also at home. The parents become an extension of Miss Postma. Besides driving their children to the center, they maintain notebooks of new words and concepts the child studies in class and attend weekly parent meetings to learn more about deafness. Most enroll in a correspondence course through the John Tracy clinic (pioneered by the wife of the late actor, Spencer Tracy, and named for their deaf son) in California, where they learn new ways of helping their children.

" 'You become enlightened because you have to. If your deaf child doesn't get good training, he hasn't a chance in the world,' says parent Bob Randall. 'We've learned a lot. For example, when people go up to a normal baby, they talk to it and coo over it. When they approach a deaf child, they are silent. He needs to be treated like any other child. Every time he looks at you, say something—so that he has something to look forward to. Miss Postma has pointed out these things to us. . . .' "

As far as speech is concerned, deafness is not the only

cause for bad speech. There is such a thing as "cleft-palate speech" also, although it may not always be caused by cleft palate. However, such a condition should not necessarily be looked upon as a sad state and the patient referred to a speech therapist without medical investigation first into the cause of the condition, according to Richard P. Jobe, M.D. and Ernest N. Kaplan, M.D., writing in the November, 1970, issue of *California Medicine* (*The Western Journal of Medicine*):

"Recent improvements in diagnosis and surgical treatment of cleft palate speech should cause insistence that . . . an intelligent prognosis and therapeutic recommendation be obtained. For some, speech therapy is the answer; for others, speech therapy alone compromises the patient's future by delaying appropriate surgical treatment and allowing the development of unbreakable bad compensatory speech habits.

"A characteristic speech pattern occurs in patients with unrepaired cleft palates and in about 20 to 30 per cent of patients who have had a repair of palate cleft. It also occurs in a significant number of persons who do not have cleft palates but have ill-defined anatomic disproportions of the palate-naso-pharynx relationship or equally ill-defined neuromuscular disorders.

"The primary cause of cleft-palate speech is the inability to close the palate-pharyngeal valve mechanism. A mobile soft palate can close against the nasopharynx and thus redirect the vocalized air stream to produce specific consonants requiring intraoral pressure. . . ." An inadequate palate-to-pharynx contact results in a recognizable hypernasality and compensatory distorted articulation that is called cleft-palate speech.

"The results of palate or phrayngeal (surgical) reconstruction are usually quite beneficial, although a minority of individuals still require speech therapy following surgery. . . ."

However, research on the ways body machinery generates speech is leading to improved therapy for patients with other physically caused speech defects, according to an article in the *Medical Tribune* of June 23, 1971:

"Thomas Hixon, Ph.D. and Ronald Netzell, Ph.D., University of Wisconsin Professors of Communicative Disorders and Rehabilitation Medicine, are using a variety of new approaches and instruments in an NIH-supported study of the

mechanics of normal and abnormal speech production at the Neurological and Rehabilitation Hospital, Madison, Wisconsin. . . .

"In studies on the effects of changing body postures on the mechanics of speech production in normal and disordered speeches, Dr. Hixon has found that gravity has a strong influence on the ability of some persons to speak.

"Some patients, such as children with cerebral palsy, show more improvement if initially treated on their backs rather than in a standing position, he discovered.

"A recent patient at the laboratory was a young girl who had difficulty speaking because her jaw remained open all the time and her tongue could not make the correct kinds of contact. Putting her jaw in an elastic band-like chin strap that goes up over the head improved her communication abilities. . . .

" 'Our main goal is improved clinical management of patients with various neurological or anatomical breakdowns in speech physiology,' Dr. Hixon explained."

Some of the other reasons for a child's speech to be distorted or slow to develop might be, perhaps, poor health in the first two years of life, inadequate teaching of speech or imitation of poor speech heard about him. Frustration can result from unrealistic standards set by a child's parents, which then lead to stuttering. Or the child of an overprotective parent who anticipates his every need may find it unnecessary to talk.

Conversely, children denied tenderness and caresses from parents may also develop speech problems. During their infancy, if they are allowed to cry for long periods of time without any attention, are given propped-up bottles frequently without anyone holding them, or if force-fed—all these can cause speech problems when the child is old enough to talk.

One of the hardest types of difficulty in speaking comes from apraxia, the inability to move the muscles of speech. Such children cannot put sounds together in a sequence, even though they have perfect hearing, and it is now believed to be caused by a genetic trait that possibly blocks the normal speech pattern. It may take years under the care of a good speech therapist to help such a child.

For information on a local hearing and speech clinic or agency, and any other material on your child's hearing or

speech problem write to The National Association of Hearing and Speech Agencies, 919 18th St., N.W., Washington, D.C. 20006.

Blindness:

"At the age of five, Jimmy is about to lose his only eye. He lost the other one after ten older brothers and sisters continually made a game of poking it, driving the vulnerable, multiple-handicapped child wild. He is now a ward of the state and his remaining eye can't be saved. . . ."

This was the beginning of a story in the *Chicago Tribune*, July 6, 1970 written by Linda Lee Landis, telling of Jimmy's bleak young life, a sordid record of neglect and abuse from his retarded mother, which took a turn for the better in February, 1970, when he was admitted to the new Eleanor Palmer Development Center for Blind Children, housed in the Goodwill Industries Building, 1643 West Ogden Avenue, Chicago, Illinois.

"Jimmy's devoted foster mother carried him there like an infant, screaming and clinging to her, terrified of everything around him.

"Now he can sit and eat by himself. He's not afraid of people any more and he can tell me where he hurts by pointing. . . ."

This Center was opened by the Lighthouse for the Blind, and a parent or guardian must accompany each child because the program is designed as a learning experience for both. The child, who usually has other handicaps besides blindness, explores the acts of daily living and learns self-discipline, while the parent learns the technique of handling a handicapped child and how to curb overprotectiveness.

"'A mother has to learn not to go to the crib of her blind child and pick him up without talking to him from the minute she enters the room,' said Miss Ethel Heeren, a blind, middle-aged staff member of the Lighthouse. 'She also has to learn not to thrust the infant into my arms without first letting him hear my voice and hold my finger.'

"When a parent and child come to the Center, leaving other children at home alone, one of the five Chicago social agencies cooperating with the center provides a baby sitter.

"'One of our goals here is to keep the family intact,' said Miss Heeren to writer Landis. 'In most cases, just the fact of

having a multiple-handicapped child wreaks havoc on family life.

" 'We also feel a blind child should be trained in a range of activity to keep his little body from becoming atrophied. My own mother used to say, 'Look at her, she's three years old. She can't see, and she can't even stand.' The normal child reaches, grabs, kicks, but the blind child is inhibited from this growth and the discovery of what his body is.

" 'These children can't meet the requirements of the public school special education program until they are toilet trained, mobile, can feed themselves, and learn some basic socialization. Parents need help with this and it's not easy to find.'

"Other family members besides parents are invited to come to the center for consultation. (Sometimes, for instance, an uninformed grandparent can ruin a child's training, by justifying leniency with, 'But, he's blind!'). . . .

"Lunch time is a communal, relaxed affair for children, mothers, and staff who sit around a long, low table on tot size chairs. One little girl looks bald from the nervous habit of pulling out her hair, rolling it into little balls and popping them into her mouth.

"Conversation is sporadic" continued writer Linda Lee Landis, in her helpful *Chicago Tribune* article, "between fits of head pounding on the wall and upset bowls of soup. Three-year-old Donald, blind, deaf, and a slow learner, sits at the table trying to feed himself, but by the time he has located his mouth with his spoon, the spoon is empty. Beside him sits his attractive 21-year-old mother. Donald's handicaps are the result of an undetected case of German measles in her second week of pregnancy. 'My mother told me I caught every childhood disease that came around, so I didn't even think about getting shots for German measles,' she said.

"Watching this mealtime scene on the other side of a one-way window is the Center's young director, Milton J. Samuelson. Intent, with sympathetic blue eyes, he talks about the children and the Center in a matter-of-fact way.

" 'There's no way of knowing how many of these children are being hidden out there in the wallpaper, getting no help, no education. . . . Illinois doesn't have a central registry yet where cases are recorded and most parents are unaware of the legislation entitling their handicapped child to a preschool education. Sometimes the parents don't speak English and have an even more difficult time getting help.

"'One set of parents told me they'd gone from clinic to doctor to clinic, looking for help on how to care for their retarded daughter. Again and again they were faced with a professional trying to break the news to them that their child was retarded. They'd accepted that fact three years ago; all they wanted was education for themselves on how to help her. . . .'"

It costs between $250 and $300 a week per child for this help for his parents and himself at the Center. The Chicago Board of Education pays up to $2,000 a year per child, an amount allotted by Illinois State Law for the private education of such children. No charge is made to the parents.

If you have a child who is blind, with or without other multiple handicaps, every state has a Crippled Child's program similar to the Illinois one. Your doctor is supposed to register a defective child (mental or physical) on the day of its birth, and it is wise when the child is brought home to check with your pediatrician as to whether he has put in this application for you. It must be signed by the doctor, not by you.

Still more fortunately, in just about every city and state in the United States, there are chapters and branches of all kinds of organizations to help a blind child. And more and more attention is being paid to the multiple-handicapped blind child. For instance, in 1966 a group of people, special educators, social workers, and medical people began to meet regularly in New York City to see what could be done to improve the situation caused by the rubella epidemic of 1964. Among these interested individuals was Dr. Louis Cooper, professor of pediatrics, New York University Medical Center, New York City, in charge of its Rubella Project, whose information on rubella-caused handicaps we quoted in the foregoing section on birth defects. He has given us permission also to quote the following from his article in *The New Outlook* about their education under his Project's supervision:

"The sharing of ideas and feelings was most valuable, because of the diversity present in the small group, which averaged about 12 people. Out of this . . . grew a number of small programs attempting to serve the needs of the multihandicapped. Our Rubella Project has been working closely with all of the new programs in metropolitan New York which have been stimulated by the impact of the 1964 epi-

demic. However, we have been most directly involved in a joint venture with the Office of Special Education of the New York City Board of Education. In 1967 Mrs. Rose Lindner, assistant principal, and Dr. John Harrington, principal of the School for Language and Impaired Hearing, and Mrs. Frances Savage, director of the Bureau for Visually Handicapped, said that they would be willing to provide a teacher of the deaf and a teacher of the blind if we could supply appropriate space, medical personnel, and a few odds and ends. We would attempt to provide services for children who could not be managed by any single discipline alone. We found a ward recently renovated by Dr. Howard Rusk temporarily empty in Bellevue Hospital. Three hardy teachers, a teacher of the deaf, of the blind, and of the brain-injured came to work there. During the past two years it has become quite clear that a multidisciplinary team can provide services to children who could not be cared for by any single member of that team alone.

"The role of paraprofessionals in any school for the handicapped is critical and we are fortunate to have health aides donated by another New York City bureau, the Bureau for Handicapped Children of the Department of Health. An occupational therapist has been supplied by the Children's Recreation Service of Bellevue Hospital, and a music therapist by the Musicians' Emergency Fund. The medical and social service support comes from our Rubella Project. The staffing ratio allows much one-to-one attention of the kind necessary with severely handicapped young children. Four of the original 20 children progressed enough that they became acceptable for educational programs.

"Our facility is far from fancy. The furniture was hospital discards. Teaching equipment and large toys came from many sources. The fact that the ward has a terrace was an added benefit because it facilitated outside play.

"Parent counselling is a critical aspect of any early intervention program. Unfortunately, I do not know a single individual who has all the skills necessary to provide proper counselling to all of the parents in a single classroom for multihandicapped rubella children. There are just too many skills required. Perhaps the same kind of approach used to create multidisciplinary medical teams could be used to provide adequate family counseling. That is, take a good psychologist, one who has an interest and knowledge of

handicapped children, a social worker with similar experience, special educators from various disciplines, and physicians, and use them as a team to provide backup for the person that relates to the individual mother. Giving the family counselor such resources may allow a social worker or teacher to guide the family of a multihandicapped child whom she would be otherwise unable to manage.

"The most important lesson we have learned from the school experience is that educators from various disciplines, doctors, social workers, and psychologists do have a good deal in common; can work in the same room together and, in fact, *must* work in the same room together if they want to provide care for the multihandicapped child. . . ."

Although there is still ignorance about the needs of a child who is only blind and has no other handicaps, and there are the occasional cases of downright cruelty, such as the blinding of little Jimmy by his ten other brothers and sisters and a retarded mother, all of whom must have been devoid of any normal sensitivities, there are many enlightened people—including parents and relatives of such blind children—who have no desire to hide or hurt them, and want only to help them become educable so that they can become adults with a reasonable assurance of earning their own living, if necessary. Many people who were born blind do not miss a faculty they never had, and go on to good vocational and other lives without feeling at all sorry for themselves. Blind children learn to compensate for the loss of their sight by using other senses more completely than even normal children do. And the best time to teach them, educational experts for the blind will tell you, is during their infancy and preschool years, before repeated frustrations undermine the child's confidence or make the parents over-protective.

Instead of trying to make a blind child describe something he cannot see, educators teach him to use the items necessary for his well-being—from being able to pull out his own chair at the table, seat himself on it, pick up and eat with utensils, to handling all kinds of equipment in the kitchen, bathroom, bedroom and even outdoors. It has been found that blind children seldom fall from jungle gyms in playgrounds, and if they are taught to use rope guides in games, they are even able to compete in foot-racing and other sporting events.

Two schools in Los Angeles, California have this method down to a science—The Foundation for the Junior Blind and

The Blind Children's Center. The first one takes blind children of all ages, but the second takes only children up to six years. Most other eye groups are not schools so much as research centers. Your doctor, however, can advise you of schools, organizations and facilities to help your blind child become a well adjusted person.

Music is often an added accompaniment to the training both at home and in school of unsighted youngsters, but wherever blind children are taught outside the home, parents are seldom allowed to be actual aides, only observers, because their concern for their own child might affect other children and parents.

It is difficult not to be concerned for your own child, but once you have gotten over the sorrow and shock of having one who cannot see, you must determine *not* to substitute for his eyes, except where necessary. He must learn to use his other senses, and use them well, and while he will need your love, your care, your sharing of experiences *with* him, he must never expect you to live life *for* him.

Should you feel it is too soon, and your child too young, to reach out for education and other outside facilities for him yet, we suggest that you write to the American Foundation for the Blind, 15 West 16th Street, New York, New York 10011, who can tell you about blindness and their many programs for helping blind people to help themselves.

Home Care of Birth-Defect Children:

Since the care of a birth-defect child is usually one you must accept for many years, apart from whatever hospital or clinic or school visits he must make, it is best to know from the beginning about ways in which you can make this care of your child less difficult for yourself, your family members, and the patient.

The *Journal of the National Association of Retail Druggists* in the July 6, 1970, issue, had an article of such tips, and we appreciate its editor, Louis E. Kazin, permitting us to excerpt these ideas for you:

"Plan the sickroom for the patient's comfort" (this, of course, would mean after the period of time when the newborn infant remains in his crib in your room), "and for your convenience. Try to have the sickroom as quiet, pleasant, and

well ventilated as possible. It should be as near the bathroom and your working areas as possible.

"For a long-term convalescence, rent or buy a hospital bed. . . . " Again, this would be at the time your baby outgrows his crib, of course, and requires at least a junior-size bed, possibly by the age of two or three. "It will be a great boon for him to have his bed adjust for leg and head elevations. You will not have the back strain which is inevitable if the bed is low and you will have to stoop in order to take proper care of the patient's needs. You may also rent or buy the bed-rails for the hospital bed which are required when the patient is a small, restless child. . . .

"If your patient is incontinent (uncontrolled wetting) or is vomiting, it is wise to use a good quality plastic mattress cover and draw-sheet for the complete protection of the bed. Pillow covers of rayon knit plastic are also available and may be used for many years as they are completely washable. Incontinent pants and liners are available in all sizes. Snap-in liners of triple layer flannel assures round the clock protection for incontinence, once a child has grown out of the diaper stage.

"On the side of the bed, be sure to pin a small paper bag. In this can be placed the disposable tissues you use for the child and waste material." (If he is not mentally retarded, he can do this for himself as he gets older.)

"It will be most helpful for you to have a good quality cover-all type apron to hang on the door of the patient's room. . . ." (For use when you change him or feed him or do anything that might get your regular clothes too dirty, and you do not want to take the time to completely change.)

"A good, stout backrest is invaluable in any home. They may be used throughout the years, not only for sickness but for watching television or reading in bed.

"A good bed-board, either the folding variety or just straight fibreboard, may be just what the doctor wishes in order to give the child better support. These boards may be taken on trips and placed easily under the mattress at night." (But be certain to get your doctor's confirmation that it is needed and helpful, don't just use one automatically for your child's birth problem.)

"Another addition to every household is a good bedside table. It is apparent that when the child is in the preschool

stage he should have his own books, magazines, radio, puzzles, as well as a place for his wash basin, his medications, right next to him or the bed. There are several types of such tables available—one to be placed by the bed, one on small legs that fits over the patient, or a deluxe hospital type that fits completely over the bed on rollers that may be pushed up and down at will.

"If he cannot go to the bathroom alone, then purchase such small items as bed pans, emesis basins, and wash bowls. They may be sterilized and reused for many years.

"Make sure the lighting over the bed is sufficient to avoid eyestrain. Place a bell by the bed so that you may be rung for, when necessary.

"Have your doctor or pharmacist show you how to take the temperature of your child. Handle the thermometer carefully, moistening it with cool water and soaping well. Take a piece of cotton and then rub downward with a circular motion. Rinse, soap again, and then rinse. Put it back in the case and have it always available when necessary." (This writer's suggestion is to buy three thermometers at once—one for the room where you feed him, if he is well enough to be taken out of the bedroom, one for the bedroom, and one for the bathroom in case one of the others breaks when it is needed. Also, get one that goes up to 108°F—instead of the standard 106°F. In conditions where there are sudden rising temperatures, infections, and convulsions, the febrile temperature will frequently go over the so-called high point of 105°F. And do not believe the old wives' tales that anything over that point will damage the child mentally. Our Claudia's temperature went between 107°F and 108°F many times, and she's as bright as any young woman would ever want to be.)

"You will have a heavier work schedule. This requires that you take care of your own health. Ask the doctor's opinion about getting in a Public Health Nurse" (or Visiting Nurse —and after this list of tips, we have some other ideas on that for you) "to help you with some of the more burdensome requirements of taking full care of your handicapped child.

"Be sure to keep a record of (whatever the doctor wants you to keep) concerning the child's daily pulse, temperature(s), urine and bowel movements—their color, amount and character; the food given and his reactions to it; his medicines and if he was able to tolerate them—(and) any (other)

unusual reactions you might observe (concerning his condition).

"Have a regular time for meals, medicines, rest periods, a bath, and linen change. A good back-rub, smoothing of the linen, opening the windows, fresh drinking water, checking of the bell and light, placing the bed pan and wash basin near if the child is able to use them—this is the kind of nighttime procedure that will make a child feel cared-for.

"In a small child, more frequent attention will be welcomed as the day becomes boring. Mealtime can be a most important time in (his) day. (If you have the time and can afford it financially) use an attractive tray, pretty disposable place mats and napkins, and have the food prepared with an eye to color and texture. Always have the child comb his hair, wash his teeth and hands before you bring in his food.

"Even children prefer feeding themselves, if they can. If you must feed him, please give small mouthfuls and alternate the food, speaking pleasantly of happenings of the day. Please do not place too large portions of food on the tray. It is most discouraging for a child to see a large man-size dinner in front of him when his appetite is necessarily small. Seconds may always be offered rather than too much at the outset."

Remember that many of the disease and birth defect groups can either help you borrow wheelchairs or crutches and health recovery equipment, or will guide you to places where they may be purchased for less. Your pharmacist can also be helpful in referring you to stores that sell the sickroom products you will need such as special dressings, swabs, sponges, and absorbent rolls of bandages that can be easily obtained locally, packaged for home use. For instance, Johnson & Johnson, manufacturers of many of these supplies, advise that to protect bedding and to cut laundering costs when caring for incontinent patients, disposable underpads are available for as little as seven cents apiece. Again, your pharmacist can either secure these for you or find out where you can get them.

Johnson & Johnson also suggest that you should plan your nursing time with a realistic view of how much can be accomplished each day. They have a few hints: always assemble everything you are going to need before going to your child's room to care for him; bring everything to his bedside

on a tray, so you will not have to leave for anything else until you finish. When finally you do finish, clean up immediately, and put everything in its proper place.

Since taking care of your birth-defect child is going to be a long haul, one thing you must be certain of is that you eat properly, have naps in the day when you have not slept well at night because of your child's needs, and take time out (using a friend or relative as baby-sitter) for walks, the beauty parlor, a coffee break, or just shopping. Discuss whatever problems are on your mind with such qualified people as your doctor, pharmacist, Visiting or Public Health Nurse, family counsellor, specialist in such problems, psychologist, or even your religious advisor. Don't hold these things inside of you until you feel you will burst if you don't get some advice or help.

Probably at times, you will think you are living in a vortex of one dilemma after another, one emergency after another, one hospital episode after another, but eventually you learn to accept even the worst of these episodes as routine, you learn to meet and conquer them, and you realize that loving your child as you do, you actually prefer to be the one there to help in these situations. You will learn that it is a lot better to be near your child, at home or elsewhere, than it is to wonder whether strangers are giving him the right kind of care because you sent him away from you.

That is why you must read the next listing very carefully, knowing that when the time comes, you do not have to shoulder everything alone, and that there really is someone available to help you during all kinds of emergencies.

The National Council for Homemaker-Home Health Aides Services: The National Council for Homemaker-Home Health Aides Services, Inc., 1740 Broadway, New York, New York 10019, (212) CI-55-8000, has a network of agencies all through the United States, whose main purpose is to provide homemakers—*not* domestics—to families in need of such help.

For example, besides helping in a home like yours where a handicapped child might require so much care that the stability of the family is threatened, this organization also cares for:

A family with an aging or chronically ill person or a neighbor who needs help to continue to live at home, a man whose wife's sudden illness or injury necessitates his being

absent from work to care for the children; a family where the mother's mental illness results in removal of the children from the home; a person who might be at home if appropriate home health care were available; or a mother who neglects her children because she is overwhelmed by their needs—a mother who has never been shown how to care for children and a home.

You and any other families described above need to know about Homemaker-Home Health Aides Services. The homemaker gives personal care, tends the sick, plans nutritious meals, bathes the patient, and contributes to his rehabilitation —but she is not a nurse. She comforts the disturbed child, encourages the depressed parent, teaches the retarded mother, understand the fears and frustrations of the aged—but she is not a social worker. She cares for the children, shops and cooks for the family, does the laundry, cleans the home—but she is not a domestic.

The homemaker who may help in any of these ways is a mature, trained, supervised member of a professional team. Her services are provided through a public or voluntary health or welfare agency to enable people to remain in their own homes.

The National Council provides technical assistance to homemaker agencies in such areas as recruitment and training of personnel, maintenance of standards, interpretation and promotion of the service and information about new uses of homemaking service. It works with health insurance companies, united funds, federal and state governments, foundations, and national voluntary health and welfare organizations to promote and finance homemaker service.

You must remember, however, that these homemakers are not offered on a permanent basis, but on an emergency basis, for only long enough to sustain the family or the individual until other plans can be made, or until the family or individual can do without this extra help.

If you want to find out the agency in your area which can arrange for this temporary homemaker service when you need it during a particularly hard time in caring for your birth-defect child, write to the organization at the New York address we gave at the beginning of this section, and enclose a stamped, self-addressed envelope for the information. You will receive it right away.

Miscellaneous Groups, Ideas, Books and Services: Where

there was no special care, other than diet and medication, I did not include further material in this section, other than what was in the description of the birth defect itself, such as diabetes, and others. For further information on these, there is a chapter of the Diabetes Association in every large city, and the national organization is the American Diabetes Association, Inc., 18 East 48th Street, New York, New York 10017. One of its major interests is patient education. Talking books and machines on which to use them are now available to persons with other disabilities besides blindness. The Library of Congress has extended the program to those who cannot read conventional printed materials because of physical limitations. Included in this group are persons with muscle, joint, or nerve involvements that affect strength and coordination.

As mentioned before, and important enough to mention again, every state in the United States has a Crippled Children's program. As soon as a child is born with a birth defect, be it mental, physical, pulmonary, neurological, your pediatrician is supposed to enroll the baby in this program, which helps the parents with costs of medication, therapy, clinic visits, and other problems inherent with caring for a birth-defect child. It also directs you to some of the services available through state-funded programs to help your child.

Thanks to Arnold Arnold, the top syndicated columnist on children, we are able to bring to you the information on Federal benefits for children from the U.S. Social Security Administration:

"If either working parent is unable to work for 12 months or longer because he is disabled at any time before retirement age, or when he (or she) reaches retirement age, then the other parent, and each child below the age of 18, are entitled to an additional monthly payment. Each is equal to one-half of the disabled or retired parent's Social Security benefits. There is a maximum that may be paid to each family. Therefore, if there are more than a given number of children below the age of 18 in your family, no more can be paid out to you.

"In the case of the death of either working parent, all children in the family below the age of 18 (and the mother, if she is the survivor) are entitled to monthly payments, each equal to three-quarters of the Social Security benefits due the deceased.

"If a child is over 18 (up to the age of 22), if he is a full-time student at an accredited school, and if he qualifies under the conditions described in the above paragraphs, he is entitled to insurance payment benefits identical to those for children below these ages. . . .

"The following is one of the least known Social Security provisions. It applies to all families WITH A CHILD WHO SUFFERS FROM ANY BIRTH DEFECT, HANDICAP, OR MENTAL RETARDATION, or one below the age of 18 who falls victim to a lasting injury. Such a child AND HIS MOTHER, as long as she personally cares for him, are each entitled to the same benefits as those detailed in the first two paragraphs above, subject to the same qualifications, FOR AS LONG AS THE CHILD LIVES.

"Social Security payments are, of course, dependent on the amount of money earned and deposited during the working life of a parent. They are also adjusted in the case of retirement, if the adult beneficiary has other *earned* income. But they are not affected by *unearned* income, like interest from stocks and bonds. . . ."

(Again, our sincere thanks to Mr. Arnold for permitting us to use this material for your benefit and that of your birth-defect child.)

If you have a child with a cleft palate and-or lip, the Mead Johnson laboratories (through your pharmacist) will supply you with the special *Beniflex* Nurser, the same nurser that was used in the hospital for the feeding of your baby, made especially to accommodate the lips and palate of a baby with this handicap. You can also write to Mead Johnson & Company, ATTN: Public Relations Department, Evansville, Indiana 47721 for a copy of "Your Cleft Lip and Palate Child, A Basic Guide for Parents" which is compiled by Gilbert B. Snyder, M.D., Samuel Berkowitz, D.D.S., Kenneth R. Bzoch, Ph.D., and Sylvan Stool, M.D., and sponsored by both the Florida Cleft Palate Association and Mead Johnson Laboratories. We suggest you send *two* 8 cent stamps and and 8 x 10 inch self-addressed envelope folded within your request for this mailing.

Should your child have a cardiac condition that eventually requires surgery, check with your County and State Medical Associations concerning hospitals that will do this kind of surgery without charge, in case you cannot afford it. Also, you local Heart Association will advise you and possibly

help you find either the funding or the hospital services for such surgery.

The *Dallas-Times Herald*, Dallas, Texas 75222 has a marvelous column called "From the Wheelchair" by Pat Galbreaith, which gives handy tips, and seeks them to use as well, concerning life in a wheelchair. We would suggest that you ask your local newspaper to use this column on a syndicated basis, or else ask them to use other information about birth defects, handicaps, etc., to serve you and others needing this way of being in contact with others.

If your child is crippled, and needs other kinds of care, including surgery, contact the local or state chapter of the National Society for Crippled Children and Adults, 2023 West Ogden Avenue, Chicago, Illinois 60612, which sponsors the very worthy Easter Seal Societies, of which there are hundreds throughout the United States. These are the groups which lend equipment for the handicapped, have camping programs for the children, and can help you and your child in any number of ways.

The National Osteogenesis Imperfecta Foundation, Inc., 28 Maple Drive, Hogansville, Georgia 30230 was incorporated in the summer of 1970 and already has a number of city chapters which hold regular meetings. This foundation publishes a quarterly newsletter containing the latest information about research on this crippling disease of the bones. Write them direct about whether they have a chapter near you, if your child has this problem.

A brochure about a preschool training program for deaf children up to three years of age is available free from the Lexington School for the Deaf, 26-26 75th Street, Jackson Heights, Queens, New York 11370. Parents, doctors, community agencies, and others interested in the problem of hearing impairment of infants and young children in New York State will be particularly interested. Because research and experience have demonstrated that the most effective training period occurs during the child's first three years of life, this branch of the Lexington School has been programmed to service children from birth, with its extensive counselling, diagnostic, and treatment facilities.

Several programs administered by the Department of Health, Education and Welfare have authority to support activities in the field of recreation and mental retardation.

These are included in the programs described in the following publications: "Mental Retardation Activities of the Department of Health, Education and Welfare." It's available for 50 cents per copy from the Superintendent of Documents, Washington, D.C.; "Financial Assistance Programs in Mental Retardation of the Dept. of Health, Education and Welfare," also available from the Superintendent of Documents, Washington, D.C., but for only 25 cents a copy. Four nonprofit organizations in the field of mental retardation which offer assistance are: The Kennedy Foundation, 1411 K St., N.W., Room 402, Washington, D.C. 20005; American Association on Mental Deficiency, 5201 Conn. Ave., N.W., Washington, D.C. 20015; National Association for Retarded Children, 420 Lexington Avenue, New York, N.Y. 10043, and Division on Mental Retardation, Council for Exceptional Children, 1201 16th Street, N.W., Washington, D.C. 20006.

For repair of any facial deformity—whether it be cleft lip and palate, or from an accident, or some other birth defect such as microtia, absence or near absence of the outer ear, or a malformation of the jaw, in which the chin sticks out too far, or hypertelorism, a condition in which the eyes are set too far apart—there is a special organization, The Society for the Rehabilitation of the Facially Disfigured, Inc., 550 First Avenue, New York, New York, (212) OR-9-1534, which sponsors the program of the Institute of Reconstructive Plastic Surgery of the New York University Medical Center and is a unique voluntary, nonprofit organization dedicated to the repair of injuries to the face from congenital causes or accidents or disease. The society prefers to concentrate its efforts on those disfigured by birth defects, accident, or disease, rather than on those seeking plastic surgery for cosmetic purposes.

Patients come there from all over the United States and many from abroad, but those writing for information are advised as to clinics and plastic surgery hospitals in their own areas.

If you or your doctor have an interest in the latest braces and other devices for handicapped individuals, especially children, write to the Director of Orthotics and Prosthetics at the Institution of Rehabilitation Medicine, New York University Medical Center, 400 East 34th Street, New York,

New York, which can advise you of their newest experiments and developments, and direct you to places producing these or something like them near your area.

If you have a cystic-fibrosis child, and want to take him abroad or somewhere where you worry about getting the proper voltage for your special mist medication machine, be advised that through an article appearing in the *Sunday Bulletin*, Philadelphia, Pennsylvania, we learned that Air Shields, Inc., 330 Jacksonville Road, Hatboro, Pennsylvania has Dia-Pump compressors weighing only 17 lbs. which are each equipped with a 50 cycle, 220 volt motor and can be used abroad. This firm is deeply involved in cystic fibrosis and inhalation therapy problems at the hospital level, and if you do not live in Philadelphia, perhaps by writing to Air Shields, Inc., they will tell you of a branch or affiliate of their service near where you live.

It is bad enough when a mother has to live with the various problems of a birth-defect child, but it is even worse when the father of the child is not as adaptable as the mother, such as the father mentioned in a letter to Ann Landers which we read in the *New York Mirror* of July 1, 1971, who refused to let his wife have another child, because he was afraid she might have another "deformed kid." Here is Ann Landers' marvelously effective reply: "Your husband is far more defective than the child. He needs counseling—and he needs it at once. Ask your pediatrician to suggest a therapist who is experienced in helping parents of children with congenital defects. Not only is your husband making himself miserable, but he is ruining his marriage and damaging the boy emotionally."

We include this anecdote for your use in the face of any heartless remarks by anyone you know, family or not, who refers to a birth-defect child as "deformed." Ask them if they are still living in the Dark Ages.

The most effective program for handicapped children about which we have read or been told is the Co-ordinating Council for Handicapped Children, 5512 Hyde Park Boulevard, Chicago, Illinois. They have a marvelous booklet, which costs only 25 cents, and we recommend that you send them an additional 25 cents as well for "Your Rights As Parents of a Handicapped Child." Address your order to Mrs. Charlotte Des Jardins, Co-ordinator there, who suggested to us "although the booklet explains Illinois legislation

on special education, it can be very useful to out of state residents who would like to make similar programs mandatory in their own state."

The American Association on Mental Deficiency, 5201 Connecticut Avenue, N.W., Washington, D.C. 20015 publishes a directory of residential facilities nationally for the mentally retarded, for $5.00 per copy (which includes postage).

The National Society for Crippled Children and Adults, 2023 West Ogden Avenue, Chicago, Illinois 60612 publishes at $1.50 (postage included) a directory of camps throughout the United States for the handicapped, some of which cost very little, some of which are subsidized.

The National Odd Shoe Exchange, 1415 Ocean Front, Santa Monica, California 90401 supplies individual shoes (when they have what you want) for people whose two feet are of varying sizes because of disease or crippling, or who may be missing one foot because of congenital causes or amputation. This organization was founded in 1943 by amputee Ruth Rubin Feldman, who keeps thousands of names on file and does try to match up mismates. Lifetime membership is $3.00, and includes your promise to send any odd, unworn shoes of your child's when you ask her to find you a single one of some special style. No promises made, but they do their best.

Problems with dressing your child? Regardless of his handicap, don't frustrate him with buttons, snaps, and so forth. Go to a store's sewing goods department and buy Velcro strips, available in various types of fabric, most colors, and good for any type of clothing except thick winter outwear; it's even marvelous for the fly in a pair of pants. The trick of Velcro cloth is the nearly microscopic hooks on one side of the strip, and eyes on the other, but all you do to close them is to press the strip halves together, and to open a slight tug does the trick. Just great!

The Exceptional Parent, Box 52, Newtonville, Mass. 02160 is a new magazine, unique among educational and professional publications, which aims to provide practical help for parents of children with disabilities. It deals with many topics affecting such handicapped children and covers such subjects as the role of the family, the nature and role of the various professional groups a family may contact, and the ways in which certain aids can be helpful. The magazine also

provides a way for parents to exchange ideas, share concerns and discover new approaches to common problems. It costs $6.00 a year.

Books That May Also Help You: *Caring for Your Disabled Child* by Benjamin Spock, M.D. and Marion O. Lerrigo, Ph.D., a Fawcett Crest paperback at 95 cents. An excellent hardcover book is *Living With A Mentally Retarded Child: A Primer for Parents*, by Beatrice Buckler, via Hawthorne Books at $6.95. Another outstanding hardcover book, *Teaching Your Child to Learn*: *From Birth to School Age*, is by our favorite writer-expert in the field of infant learning, Arnold Arnold, via Prentice-Hall, Inc. at $6.95. Last, an original paperback, *The Complete Directory of Medical and Health Services* by yours truly, Jessyca Russell Gaver, via Award Books at 95 cents.

4

The National Foundation—March of Dimes

What It Is and What It Tries to Do:

In all the years of being a magazine and book writer, this is the first time, in trying to interview representatives of an organization, that nobody would discuss his own work, merely that of others—such as the work of the medical people and of the volunteers.

Only by finally receiving permission to quote from "Facts —1971 March of Dimes" could I learn *some* of its background. And further on, I did the same thing with their 1971 Annual Report, "Focus on the Family," After which I received the help of a few people, who know the March of Dimes work well, with updating my material:

There are two ways to think about birth defects. One is the old way—they are just one of those tragic things that happen, nothing can be done about them. The other is the new way, The National Foundation-March of Dimes way: birth defects are the second largest killer in the U.S.; they kill or cripple at the beginning of life; they strike the most treasured, most defenseless part of our population, the nation's children, so they are a major public health problem in this country.

Enough said? Never! Not when a health problem affects a quarter of a million newborn babies annually. The National Foundation can and is doing something about birth defects. Some day it hopes to guarantee to every American child his birthright: a whole mind in a whole body.

Yes, prevention of birth defects is the goal of The National Foundation. That's what the March of Dimes is all about. But there are a lot of doubters in this country and even in the world.

When the original National Foundation for Infantile Paralysis (poliomyelitis) was founded in 1938 by Franklin Delano Roosevelt, people thought it was impossible to prevent

polio. Today, cases of polio are so rare in this country that there were only 30 cases in 1970, not the epidemics known previously.

Everybody knows (or they should know) that this magnificent achievement was the direct result of the massive assault on the polio problem mounted by the March of Dimes during the 1940's, culminating in the development of the Salk vaccine in the 1950's and of the Sabin vaccine in 1961 and 1962. And everybody should also know that Drs. Salk and Sabin, along with scores of other distinguished scientists, were backed to the limit by the Foundation.

The Foundation set out not just to take care of polio victims, but to prevent polio, and in partnership with American medical science, volunteers who helped raise the funds for this, it did the job. By 1958 the victory over polio was assured. Meanwhile, the Foundation's trustees had begun in 1952 to survey the public health situation. They decided to bring the Foundation's tremendous experience to bear on a long-neglected but extremely serious child health problem: birth defects.

It has already made important strides along this road. When it tackled the problem late in the 1950's, the very term "birth defects" was unknown. People vaguely understood that some babies die in the womb for some reason and that others are born with defects that seriously—sometimes ruinously—impair their lives. Even many doctors were only generally informed about the scope and severity of the problem.

Not so today. Knowledge and understanding of the mechanism of genetics have expanded unbelievably, as we explained in the first section of this book. In 1970, scientists succeeded in isolating the basic unit of heredity, the gene, and another group of researchers actually synthesized a gene. Science has taken the first, terribly important steps toward prevention of genetically caused birth defects. The barriers are down.

Of course, the National Foundation-March of Dimes does not claim credit for every new scientific discovery related to birth defects. It is merely proud that some of the advances have come from the laboratories of men whose work it has supported, such as the 14 recipients of the Nobel Prize who have been associated with The National Foundation, and if you wish to know who all of them are, we are sure their

public relations department will be glad to supply their names and biographical data.

My reasons for devoting this last section of this book to the work of The National Foundation-March of Dimes is because of the selflessness of their many representatives with whom I worked. Chief among them was Mrs. Camille Jackson, of their Professional Education Department, who works with the editors of much of the scientific material that goes to doctors and other health professionals on birth defects. It was she who suggested I see two wonderful doctors who are devoting their professional lives to helping birth defect children: the aforementioned Dr. Louis Cooper of the Rubella Project at New York University Medical Center, and Dr. Alfred Scherzer of the Spina Bifida Project at the Birth Defects Center of Cornell University Medical College-New York Hospital, both in New York City.

Dr. Cooper wanted me to bring the baby to his associate who specializes in spina bifida at New York University Medical Center, because the hospital in New Jersey where little Jimmy was born knew little or nothing about the treatment for this defect. But Dr. Scherzer gave me the name of the Director of the Birth Defect Center at the Babies Hospital Unit of the United Hospitals of Newark, New Jersey, because my daughter and her husband live in New Jersey.

You may find it hard to believe how the medical personnel at this Newark unit tried to save little Jimmy's life—and kept trying for the whole nine days he was there. That was when our little family decided to do something for all those connected with The National Foundation-March of Dimes who had tried to help little Jimmy. It had not cost us a penny at the Newark unit for little Jimmy's care and treatment, and we felt that his loss had been one for them as well as for us. Even those busy, wonderful medical-grantee new friends of ours kept in touch with me—Drs. Cooper and Scherzer—hoping my news of the baby might be good, encouraging my efforts to find a publisher for this book, sending me to sources of information and giving me what they could, personally.

Camille Jackson kept in touch, too, and was probably the most surprised when I arranged with her to present a gift of $2,715 collected from our friends, business associates, and family for the March of Dimes Fund. It was given on June 1, 1970. We didn't have the time to serve as regular volun-

teers, but we each felt that raising money for this organization's wonderful work was the least we could do in little Jimmy's memory. That is why I am now contributing these facts to you, also, so that you can be aware of the March of Dimes facilities and, in turn, may be able to use them to help both yourself and your birth-defect child.

When Dr. Robert A. Good won the Albert Lasker Medical Research Award in November, 1970, Dr. Virginia Apgar, vice president for medical affairs of The National Foundation (who is known, among other accomplishments, for her creation of the Apgar Score, the clinical evaluation made within 60 seconds after birth to determine the newborn's over-all condition) hailed the award to Dr. Good as one of the most significant in the 25-year history of Lasker Awards and cited Dr. Good's far-flung contributions "to understanding of immunity and its relationship to birth defects, cancer, arthritis and other human disease."

"Public contributions in the cause of health have made this work possible," Dr. Apgar explained. "Since 1960, Dr. Good's research has been supported by grants totaling more than $850,000 in March of Dimes funds alone. In that period he and his associates established the crucial role of the thymus gland as part of the immune mechanism. Almost every year, they have recognized 'new' disorders of immunity and determined the nature of the underlying defect. . . ."

(Dr. Robert Good, by the way, is the pioneer mentioned earlier in this book for his method of transplanting bone marrow from a healthy donor to a child, to make it possible for victims of a rare genetic ailment to remain alive as healthy human beings.)

Finding the answers to birth-defect problems offers even more important benefits than scientific achievement. Determining the causes of a defect is a major step towards primary prevention. One way is by supporting basic and clinical research, trying to find out what goes wrong when birth defects occur—and why. More than a thousand different kinds of birth defects have thus far been identified. Now—how to prevent them?

Such research—and that of all human illness—begins with the basic approaches taken at The Salk Institute. Here the focus is on investigation into the basic principles of biologic functions: how do they work properly and how do they go

wrong? Through 1970, the National Foundation has invested $22,000,000 in The Salk Institute, located in San Diego, California, and in 1970 granted $2,688,394.

The National Foundation supports both basic and clinical research at institutions across the nation. In these programs many scientists seek solutions to a number of specific problems related to birth defects.

The Foundation supported 12 Birth Defects Research Projects in fiscal year 1970. These studies are usually conducted solely in the laboratory. There were $959,173 in grant funds for this purpose in effect during this period.

There were also 14 Birth Defects Research Centers in fiscal year 1970 with $655,905 in grant funds in effect. Patients are usually studied at the research centers, but because of the special nature of the work, medical services are not regularly provided the way they are at the Birth Defects Centers themselves, where hospital and other care is given to birth-defect babies.

Since The Salk Institute began operations in 1963, it has grown from 12 members to some 250. Among distinguished additions to the staff in 1970 was Dr. Roger Guillemin. Formerly a professor of physiology and director of the neuroendocrinology laboratories at Baylor University College of Medicine, he is now a Resident Fellow at the Institute.

Dr. Guillemin will continue his studies on the purification, isolation, and synthesis of chemical factors in the brain which regulate production of hormones by the pituitary gland to control many important body functions. One of Dr. Guillemin's special concerns is what he terms "the birth defect of a child who is born where and when he is not wanted, where and when he cannot be properly fed and raised and educated." This concern leads him to special interest in the field of fertility control. He will play a major role in the Institute's newly initiated program in reproductive biology.

Expansion in staff and in new areas of research, such as neurobiology, continues at the Institute. Within the next five years, the Institute will reach its planned maximum staff number of 450.

A wide variety of problems related to birth defects is under study in Foundation-supported Birth Defects Research Programs across the country. For example, three research teams are focusing on problems related to spina bifida and

hydrocephalus. At the Albert Einstein College of Medicine in New York City, Dr. Kenneth Shulman and his colleagues are trying to find the best methods for treating the child born with this defect and how to judge his potential for improvement.

The primary concerns of Dr. David Karzon at Vanderbilt University in Nashville are how spina bifida patients respond to surgery and how brain and blood stream infections can be prevented in patients with hydrocephalus. (This particilar research was supported by the National Foundation until February, 1971, and is now presumably supported by the University itself.)

Dr. David B. Shurtleff and his team at the University of Washington in Seattle are investigating several questions, including: What is the best drug treatment for these patients when surgery is delayed or unsafe, and how can water pressure inside the brain be gauged with scientific reliability?

Other research programs are more basic. "What turns genes on and off?" That question is asked by Dr. Vincent G. Allfrey of Rockefeller University in New York. At Massachusetts General Hospital in Boston, Dr. Jerome Gross is studying how organs develop during the embryonic period and how lower animals regenerate lost body parts.

The effects of thyroid hormone deficiency on the development of nerve cells and connections in the central nervous system are being examined by Dr. David Bodian at the Johns Hopkins University in Baltimore.

The activities of Dr. Victor A. McKusick, who has been associated with The National Foundation for many years, have gained world renown. The Birth Defects Research Center he directs at Johns Hopkins concentrates on several groups of abnormalities, including hereditary skeletal defects, absence of limbs, congenital deafness, and dwarfism.

Dr. McKusick and his staff have also developed a semicomputerized Family Oriented Medical Records System (FOMERS).

"The objective of FOMERS is preventive medicine, in relation to serious complications of certain hereditary diseases, through proper follow-up of persons known to be at risk because of their positions in their families," Dr. McKusick explains. He expects the system to assure prompt, correct diagnosis and proper treatment, to detect and eradicate life-threatening complications when they arise, to sug-

gest effective prophylactic measures and to avoid the birth of offspring with serious chromosomal aberrations.

Dr. McKusick considers this system to be "an extension of the physician-patient relationship to a physician-family relationship" which he feels will "substitute confidence for anxiety" within the family.

The Division of Medical Genetics and its clinic at Johns Hopkins Hospital is one of the world's foremost research centers in hereditary disease. Its overall emphasis has been to characterize precisely by clinical, laboratory, and family studies many of the rare and obscure birth defects in order to achieve a more rational classification of these diseases. Dr. McKusick has taken a leading role in this work.

He has also served as chairman of four conferences on the clinical delineation of birth defects, sponsored by The National Foundation. The first meeting in 1968 explored malformation syndromes, limb anomalies, skeletal dysplasias, and phenotypic aspects of chromosomal aberrations. At the 1969 conference, defects of the central nervous system, eyes, ears and muscles were discussed. At the June 1970 conference the defects examined related to the endocrine system, skin, and teeth and other oral structures. At the 1971 conference the subjects were the cardiovascular system, blood, and the gastro-intestinal tract, including the pancreas and liver.

Dr. Daniel Bergman, Vice President of professional Education for The National Foundation, acted as editorial adviser, with Dr. McKusick as scientific editor, when the Foundation published the papers presented at the first conference in 1969, and those from the second and third conferences in 1971.

Akin to Dr. McKusick's work in the delineation of birth defects are the studies of Dr. John M. Opitz at the Birth Defects Research Center at the University of Wisconsin Medical School. This center is world-famois for its work on rare congenital syndromes. Dr. Opitz and his associates have so far described 12 new syndromes and are working to delineate many more from accumulated clinical and genetic data on the many patients and their families who have been examined at the center. In particular, their studies on the developmental and anatomical aspects of genetic malformation or retardation syndromes have led to new findings of fundamental importance to human genetics and developmental biology.

Both Dr. Opitz and Dr. McKusick also serve as regular consultants to the March of Dimes-supported Syndrome Identification Service at the Boston Floating Hospital. Doctors from all over the world use this service to obtain diagnostic and treatment information for their patients with birth defects.

Scientists and physicians from many countries gathered at the Hague, Netherlands in September, 1969, for the Third International Conference on Congenital Malformations. The six-day meeting, sponsored by the National Foundation, drew more than 1,000 specialists in medicine and biology to this largest scientific meeting ever held on the topic of birth defects.

Two hundred participants from 29 countries discussed every aspect of birth defects, from known causes to means of prevention and treatment, in formal papers, in discussion, and "free communications" sessions. Leading geneticists, biochemists, pediatricians, surgeons, immunologists, pharmacologists, and epidemiologists contributed their specialized expertise to the common pool of knowledge.

Commenting on the conference when it ended, Basil O'Connor, president of the Foundation, said: "Now we can report the beginnings of achievement in a world-wide effort of science to improve the quality of human life at birth. It will not be easily or quickly accomplished, but it is within reach."

Another conference sponsored by the Foundation in 1970 was a symposium on fetal malnutrition held in January in New York. Dr. Myron Winick, director of the Foundation's Birth Defects Research Center at Cornell University Medical College, was chairman of the meeting. Although part of Dr. Winick's research program involves congenital defects of the kidney and the genito-urinary tract, he is also intensively studying the causes of prenatal growth retardation, particularly the effect of a mother's malnutrition on fetal development.

The role of malnutrition in pregnant animals and the deleterious effects it has on brain development in their offspring has been well established. Dr. Winick and his co-workers are investigating whether this is also true in man.

"Understanding the role of maternal and infant nutrition in brain development is important to those of us concerned with the prevention of birth defects," Dr. Winick says.

"Subtle problems such as mental deficiency may not be detectable at birth, but when they do become apparent they are no less handicapping than some of the more obvious malformations."

Whether a defect is obvious or subtle, certainly it is undesirable. All of these National Foundation-supported research programs have a common goal—giving every child the opportunity to develop his fullest potential.

After a family recovers from the initial shock of having a child born with a birth defect, after the parents have learned what can be done for the child and how to cope with his problems, they begin to think about the future. They are obsessed with the question: "Will it happen again?"

Too often in the past, it was impossible to provide an answer. Today, with increasing frequency, National Foundation grantees and others are providing answers through genetic counselling (explained previously in this book). Such counselling is best done as a multidisciplinary endeavor with knowledge drawn from the specialties of genetics, internal medicine, pediatrics, obstetrics, and gynecology.

At Cardinal Glennon Memorial Hospital for Children in St. Louis, Missouri, for example, couselling is provided to parents by the Center's associate director, Dr. Patrician Monteleone. She is also director of the hospital's genetic clinic and assistant professor of pediatrics at the St. Louis University School of Medicine.

One of the most useful studies in this procedure is the preparation of a karyotype. Cells from the person or fetus under study are taken and cultured. Then the chromosomes of one cell are photographed under the microscope. The images of the individual chromosomes are then numbered and arranged in a specific order according to size and shape. The resulting karyotype will show scientists such anomalies as missing or extra chromosomes, deleted chromosome material, breaks in chromosomes, or translocation of genetic material.

For genetic defects in which the "mode of inheritance" is known, the counsellor can often make firm predictions. But parents need more than just mathematical probabilities to reach a decision. Dr. Monteleone doesn't think "it is the responsibility or the duty of the genetic counsellor to recommend or make decisions" for the parents, but she tries to make them aware of the "many other factors involving

psychological, sociological, economic, and emotional aspects."

Dr. Lair Jackson, director of the division of medical genetics at Jefferson Medical College in Philadelphia, Pennsylvania, tries to explain the problem in terms of a specific family. As director of The National Foundation's Birth Defects Center there, he met with a couple from central Pennsylvania who had already lost two children afflicted with Werdnig-Hoffman disease. The first child lived 18 months; the second, 15 months. Dr. Jackson told the parents that the risk of recurrence for each subsequent child was one in four. But other factors had to be considered.

"Knowing what mothers are like, it might be that having another child whose diagnosis could be immediately apparent to the mother would be a great strain," says Dr. Jackson. "Another child that she would have to watch die might be very much more than her emotional make-up could stand at all. In that case, the risk of one in four would become something bigger than she would want to face."

Conversations with the couple showed this to be precisely the wife's feelings. The husband was willing to face the risk, but realized during their meeting with Dr. Jackson that to do so, he would be jeopardizing his wife's mental health. The couple chose to adopt children instead.

Another program which reaches patients over a wide area is The National Foundation's Prenatal Birth Defects Prevention Center at The Johns Hopkins Hospital in Baltimore, Maryland. The Center entered full-scale operations in June, 1969, and has since received more than 300 inquiries from across the nation. Dr. Richard Heller, co-director, says that its purpose is "to encourage parents to have children," parents who otherwise might not have children for fear of their being afflicted with birth defects.

Dr. Robert E. Cooke, chairman of pediatrics and co-director of the Center, explains further. "If a family has a severe disorder, it has been our experience that many times the parents won't risk having another child. So they go through life with one seriously handicapped child. If they are given assurance by amniocentesis" (which procedure we have explained on page 34) "that the child is going to be normal, then they'll go ahead."

In addition, the Center also sees pregnant women faced

with the problem of congenital rubella and other infectious diseases, and is studying the problem of drugs and their possible teratogenic effects.

Four-fifths of the women seen at the Center during the first year did not require amniocentesis. And of those patients who did, only two learned that the fetus was definitely defective and chose to terminate their pregnancies. Follow-up after a diagnosis has been made is an important part of the Center's function.

"If the diagnosis is a healthy baby, we await the termination of pregnancy and then examine the baby. So far, all of our babies have been normal," says Dr. Heller, sounding like the proud papa of all of them.

Both Drs. Cooke and Heller—as well as many other physicians—feel that follow-up should extend beyond the immediate child involved. For instance, Dr. Cooke saw a child over 20 years ago whom he diagnosed as suffering from galactosemia. The child was mentally retarded. Recently, he received a letter from that patient's sister who is living in Florida with her husband. They were thinking about having children and wanted to know whether they would have a baby with the disease, which is inherited recessively.

Shortly thereafter, the couple visited the Center for tests. The wife did prove to be a carrier, but the husband was not. "So they won't have any galactosemic babies, although they may have babies who are carriers," says Dr. Cooke.

These doctors all look forward to the development of more centers with laboratories and scientists capable of providing effective genetic counselling for parents.

"I think the public is ready to accept this information and deserves to have it," says Dr. Heller. "Certainly, every baby deserves a healthy life."

Now that science has learned how to diagnose some defects *in utero* (meaning in the uterus), the next step is to learn how to correct the defect before it has an opportunity to diminish the life potential of the fetus.

The answer to that, all scientists involved hope, would be genetic engineering, since the gene is the basic unit of heredity. Some current research is geared toward developing the ability to change, at the most basic level, the effects of specific genes inherited from one or both parents, to allow the correction of a genetic defect before it can damage a fetus, but

most experts believe the answers will be hard to find and will take a long time to discover.

What Families Can Do to Help:

When families and other concerned members of the community learn how much still remains to be done in the fight against birth defects, many ask what they can do to help.

Many examples of concerned individuals are given in the 1970 Annual Report of The National Foundation-March of Dimes, "Focus on the Family."

For instance, Donna Fry of Salt Lake City was noted as having found one answer. She is a March of Dimes service volunteer in the Birth Defects Center at Primary Children's Hospital. Mrs. Fry knows personally of the tragedy of birth defects. Her eighth child was born with a defective heart and died in 1958, the same year The National Foundation took up the fight against birth defects. Although she is a grandmother now and still has four of her children at home, Mrs. Fry has been helping at the Center since 1968. At 8:30 a.m., three Fridays a month, she begins a busy morning. Usually, about 25 patients come to each clinic. Mrs. Fry checks in patients, records their height, weight, and temperatures, takes urine specimens, and takes each family to the proper room to see their doctors. She is one of two March of Dimes service volunteers who attend every clinic session. Mary Lambert, clinic nurse, says, "it would be impossible to manage clinics of this size if we didn't have these two women to help us each time."

While there are many people like Donna Fry who individually volunteer their services in the Birth Defects Centers, there are literally hundreds of other men and women who have joined together in groups to help their committees with March of Dimes programs. Promoting prenatal care and rubella vaccination are two areas in which March of Dimes public health education programs have been highly effective. These programs are designed to assure that the benefits of medical research are applied in the community. March of Dimes volunteers work together with health departments, medical societies, and other cooperating agencies and organizations in hundreds of communities to set up programs of education and action to assure that every pregnant woman

receives prenatal care and that every child between age one and 12 receives a rubella vaccination.

The need for prenatal care is well documented in medical literature. The prematurity rate is at least twice as high among woman who have had little or no prenatal care than among those who have been under good medical supervision. A significantly higher rate of birth defects and infant deaths is associated with prematurity.

Yet far too many women in the United States see a doctor only when they enter a hospital in labor. In a 1967 study, for example, the medical advisory committee of The National Foundation's Genesee County Chapter found that a large number of women delivering at the city hospital in Flint, Michigan had never had prenatal care. After mobilizing community forces, the chapter opened its free clinic for prenatal care in an inner city area that year. Mrs. Avorita Windiate, executive secretary of the chapter fondly remembers the opening: "Our desire to create a warm friendly atmosphere developed immediately and the patients were more than appreciative.

"It was only a matter of a few weeks before the City Health Department decided to provide a health nurse each clinic morning and her discussions and films on nutrition and health problems soon became a standard pattern in the clinic. At the same time, the C. S. Mott Infant and Maternal Health program sent one of their top nurses to teach patients proper exercise and breathing. The city health department also provides all of our lab work free of charge."

Headed by Dr. William Roberson, a local obstetrician, the clinic sees about 35 patients each Tuesday in its four examining rooms. The medical community has been so impressed with the clinic that it was incorporated into the Hurley Hospital teaching program in 1970. The hospital's chief resident and three student nurses attend each clinic with Dr. Roberson.

A prenatal care program with a different slant is in operation in Atlanta, Georgia. The B'nai B'rith Women, the local March of Dimes chapter, the Atlanta Council of Catholic Women, and the Service Guild all provide volunteers for their Operation Stork project at Grady Memorial Hospital. The project is only one part of "Better Infant Births," a program dedicated to improving the quality of babies born in Georgia.

A description of some typical hospital scenes show the functions these women perform:

In a small room on an upper floor, a cheerful volunteer is teaching four young women to crochet. They are working quietly, gossiping among themselves, discussing their pregnancies, for all are high risk maternity patients.

In a play area adjacent to one of the clinics, another volunteer has her hands full of toys and children. She sits on a dimunitive chair, low enough so that even a two-year-old can crawl into her lap while his mother is seeing the doctor.

In one of the consulting rooms, another volunteer listens while a young pregnant woman tells about herself. The young woman asked to be interviewed and have her personal history taken by an Operation Stork volunteer. Her friends had told her that the ladies are easy to talk to and will listen, as long as you want, to your questions.

In the obstetrics clinic, other volunteers are helping patients plan menus for their families, especially in view of the recent medical emphasis on good nutrition during pregnancy. A big consideration is how best to use the surplus commodity foods which most of these patients receive.

Most of the Operation Stork volunteers are busy women with homes and children of their own to care for. But they are able to provide that essential which an overworked hospital staff frequently cannot make time for: loving interest and sympathy.

The ability of March of Dimes volunteers to mobilize other sectors of this community is vital to so many of their prenatal care programs. It has also proven invaluable during their campaign to promote rubella vaccination. The National Foundation was one of the first voluntary health organizations to recognize the importance of taking immediate action to prevent another rubella epidemic.

The last major epidemic in the United States occurred in 1964-65. Thousands of pregnant women contracted rubella, often from their own children. Rubella epidemics occur in a six-to-nine year cycle. Until the licensing of a rubella vaccine in June, 1969, nobody was able to prevent the thousands of abnormal pregnancies and births resulting from the 1964 one. But during 1970, the March of Dimes Chapters have been involved in most public vaccination programs conducted in the United States. In some areas, March of Dimes volunteers were the catalysts that sparked these

programs. Their involvement has covered a wide range of activities, depending upon community needs. They provide both lay and medical volunteer manpower in vaccination clinics; promote public health education concerning the need for funds to make the vaccine available; help obtain community and other public funds for vaccine purchase; furnish transportation to clinics for families without care, and assist health departments by acting as the coordinating agency for rubella vaccination programs.

March of Dimes chapters have been involved in rubella vaccination campaigns in almost every state in the union. Over 7.5 million copies of its educational pamplet, "Rubella Robs the Cradle" (including some 100,000 in Spanish), have been distributed. And about 24 million children were vaccinated in public school health programs by June 1, 1971, a major achievement. This comprises 50 per cent of the target population, but the other half are preschoolers and are therefore the hardest to reach. However, March of Dimes chapters are expanding their rubella campaigns further in the future.

Public health education and action activities are vital if the means to prevent birth defects—such as the rubella vaccine, prenatal care, and the new preventive for Rh blood disease—are to be applied in the community. Foundation volunteers have shown that their community efforts do work.

PREVENTION THROUGH TREATMENT, RESEARCH AND EDUCATION:

Although we have already discussed generally that The National Foundation-March of Dimes does give grants for research, has Research Centers and has Medical Service Centers, these were discussed in relationship to the work of individuals, as well as volunteers. Now, let's take them collectively, to get a better idea of the scope of this entire organization.

One in ten American families knows the tragedy of a child born less than perfect. A birth defect doesn't happen to just *one* person—it happens to a whole family, creating severe emotional social, and financial strains as a result of one member's handicap. It thus becomes a *family* with a handicap.

The National Foundation is concerned with the problems of these families as well as with the prevention of birth defects. The concept of team care for a patient evolved during the years that the Foundation was fighting polio. This pattern of comprehensive, coordinated care has been continued and expanded in its Birth Defects Centers. It now includes care for the family as a whole. Genetic counsellors, psychiatrists, and social workers have become important members of many medical teams to provide answers to questions an afflicted family asks.

Children born with birth defects need comprehensive diagnosis and treatment. A network of March of Dimes Birth Defects Medical Service Centers provides diagnosis and treatment by teams of medical experts for such children. They are then given long-term rehabilitation, if necessary. Some Birth Defects Research Centers concentrate on specialized research to uncover the causes of birth defects and means of prevention.

The National Foundation Medical program attempts to raise the level of care for all birth defect victims by:

(1) Setting a new example of quality care.
(2) Developing new knowledge through research.
(3) Teaching students of medicine, nursing, and other health professions.
(4) Making new knowledge available to doctors and other health professionals in their institutions and in private practice.

Many of these Birth Defects Centers are connected either with top medical centers or medical schools, and special allocations are also made for patient aid, professional personnel, or equipment when a special need exists.

Each Research Center concentrates on a specific area or type of birth defect. These Centers conduct scientific research to learn more about the causes of birth defects and means of prevention, they seek ways to provide earlier and more accurate diagnosis, and they develop improved medical, surgical, and rehabilitation techniques. They are also supported by grants from The National Foundation headquarters.

The Foundation's Medical Service Centers provide diagnosis and medical consultation by specialists as well as comprehensive care for both inpatients and outpatients. In

many Centers accredited teaching programs train medical students, physicians, nurses, and other paramedical personnel.

These Centers bring together specialists from many fields to consult on how best to deal with all the problems of a child born with defects. The team approach eliminates the confusion and frustration that comes when a family must see each specialist in a separate place. It means that the skills of a pediatrician, neurologist, orthopedist, surgeon, psychologist, physical therapist, and other experts may be focused on a single patient in a single visit. There are three other important members of this team: the physician who has referred the child to the Center and the parents who must maintain care at home. Many Centers also provide services for the entire family, such as genetic counseling.

Medical Service Centers are financed directly by grants from a chapter or group of chapters of *The National Foundation* to medical centers or teaching hospitals.

National Foundation chapters also help birth defect patients by using March of Dimes funds for special local needs not covered by existing community resources. Upon recommendation of the chapter's medical advisory committee and with approval of national headquarters, special allocations of funds may be used to:

(1) Initiate or improve birth defect services in selected hospitals or other health agencies.

(2) Provide professionals, including medical social workers, physical therapists, laboratory assistants, speech and hearing therapists, where needed.

(3) Purchase special equipment essential to proper diagnosis or treatment of birth defects.

(4) Cover costs of short-term hospital care for diagnostic purposes for children who are not eligible for help from other sources.

Education—both for the public and for medical professionals—is a major force used by The National Foundation in preventing birth defects. The recent licensing of rubella vaccine is a case in point: developing the vaccines was only half the job. Getting people to understand their importance and to use them is the other half.

Genetic counselling is another area in which education has become increasingly important. For example, physicians who are aware of the new tests being developed for detecting car-

riers of abnormal genes can suggest that a patient with a family history of birth defects can see a geneticist for tests and counselling, because The National Foundation works hard to keep physicians up to date on this kind of information, as part of its education campaign.

The National Foundation-March of Dimes steadily contributes to the accumulation of information about birth defects and makes this knowledge readily accessible to medical schools, physicians and other professional personnel. The Foundation publishes articles by leading scientists in the field, of which the collection includes:

1. Reprints Series—complete articles of special merit in the field of birth defects from United States and foreign journals. More than 164 papers are now offered, and requests for over one million copies have been filled.

2. Original Articles Series—these original papers by authorities on birth defects are published only by The National Foundation. Now in its seventh year of publication, this series has grown to 32 books.

Scientists and physicians specializing in birth defects meet at local, national, and international conferences sponsored by The National Foundation, as previously mentioned earlier in this section.

Professional films are provided for use in medical schools and medical meetings. For example, in 1970, a series of eight films on medical genetics was produced under the supervision of Dr. Victor A. McKusick of Johns Hopkins University School of Medicine. Technical exhibits, illustrated booklets, slide presentations, and other materials are available for professional gatherings, public health meetings, and at birth defects symposia.

Until two statewide programs supported by March of Dimes grants went into effect recently, there was no central clearing house anywhere in the country that listed all the facilities in a given area for those children who were not born perfect. Now such information is available from the Massachusetts State Department of Health, and at the University of Alabama Medical Center in Birmingham. These two information centers were designed as pilot programs whose main functions are to indicate paths for public health departments.

And that is exactly what has occurred. In January 1970, the Massachusetts State Department of Health assumed responsibility for the Boston information and referral service.

Pilot programs of this sort, which are not meant to fill the whole need that exists but instead to point the way for government health agencies, are one of the ways in which The National Foundation works most effectively.

The National Foundation conducts continuing campaigns of education on Rh disease and rubella, with special press kits that include press releases, television film spots, editorials, and radio announcements in Spanish and English. A film on rubella entitled "Fight Against Time" is also available for use in the continuing vaccination campaign.

Some of the other means of public education used to bring better understanding of the problems of congenital disease to the American public are:

Film Strip—"More Than Love" is designed for classroom use in senior high schools, emphasizing that preparation for parenthood begins in the teens, and highlighting the value of prenatal care in preventing birth defects.

Films—Educational films about birth defects and individual Centers are available on loan to TV stations, organizations, schools, and for special meetings or symposia.

Slide Presentations—Local speakers may order scripts and slides to illustrate March of Dimes efforts against birth defects at Centers, through research and prenatal care.

Publications—Leaflets, booklets, posters, newspaper, and magazine articles are produced for mass distribution or for specific groups such as families of children with birth defects, expectant parents, service groups, teen-agers and students in health education classes, as well as March of Dimes volunteers.

Exhibits—Educational displays at professional meetings, hospitals, health fairs, libraries, store windows, health departments, and so forth, depict the problems of birth defects and the importance of prenatal care.

Speakers' Bureau—Chapters arrange for informative speeches on birth defects, prenatal care, and other March of Dimes programs for meetings of local organizations. Members of Birth Defects Centers' medical staffs often address community groups as well as professional audiences.

Seminars or Symposia—Educational meetings for medical, paramedical, and general audiences are highly informative. Many local and state birth defects symposia have become annual events.

Where There's Help . . . There's Hope

A pamphlet written by mothers of children with the birth defects of myelomeningocele and hydrocephalus (all disorders related to spina bifida, by the way) was published by the Community Services Department of the Greater New York Chapter of The National Foundation-March of Dimes, and is reprinted here in part in the hope that its wonderful advice will inspire other mothers of children with other birth defects to bring out similar booklets:

"We decided to write this pamphlet in order to share our experiences with others. We hope it will be helpful to mothers of children with birth defects. . . .

"It is intended to ease their confusions, to help them feel less alone with their problems, and help to give them a better understanding of their children's medical status.

"We have included some references to our own emotional reactions because we have found that certain feelings are common to us all.

"We have also added ideas derived among ourselves on how to cope with the social, emotional, and practical situations which confront us and our families.

"Our discussions have revealed that, even though our children were given the same diagnosis, their early care and treatment varied widely. Advice and attitudes given to us as parents also varied widely.

"We have included information and experiences . . . which we hope may help the professionals who deal with children and families like ours. . . .

"Although (the medical terms) definitions, explained in lay terms, seem simple, some parents have been given only the medical terms without explanation after the birth of their child. The terms had different meanings for many of us. For example, when some mothers were confronted with the terms 'hydrocephalus,' or 'water on the brain,' they immediately thought their children would be severely retarded and have no hope of being physically normal.

"The term 'myelomeningocele' might not have been frightening, because it was so unfamiliar. But in some cases, a well-meaning attempt to oversimplify matters by telling a mother that her child has a 'hole in the back' had the opposite effect. Taken literally, the expression brought terrifying images to the parents' minds. On the other hand,

those who were given a clear description of the situation found that facing reality was far less difficult than trying to imagine what it was.

"As mothers who had given birth to handicapped children, we were confronted with real emotional crises. At first, there was shock and disbelief, evidenced by the thought, 'How could this happen to me?'

"Then guilt was uppermost in our minds, because we believed we were in some way responsible. Some of us suspected that some medication taken during pregnancy might have caused the defect. Others thought an injury or heavy lifting might have been the cause. One mother even thought that because she had cared for a myelomeningocele child while pregnant, she had caused her own youngster to have the same problem. Others felt inadequate as women, because they had failed to give birth to perfect babies.

"Many of us were beset with loneliness, despair, and fear. What would become of our children? How would this affect our families? How could we face the mounting expense of medical treatment?

"Because of the surgical problems we were separated from our children early in their lives. Did this mean we would never develop a normal mother-child relationship . . . ? We wanted to talk, comfort, love, and hold our children. This seemed most urgent to those of us who had been told that our children faced the possibility of early death.

"Our fears posed serious problems which threatened marital relationships and led to over-protection of the afflicted children. Some thought their child's future looked so frightening that it might be better if the child would not live.

"Realizing our emotional needs, we have searched for ways to deal with these difficulties. Candid discussions with each other led to a clearer understanding of the facts and enabled us first to more easily recognize and handle our problems.

"For example, some of us could face our guilt feelings when we began to understand that the exact cause of myelomeningocele is unknown. The damage occurs in the early weeks of pregnancy, a time when most women are not even aware they are pregnant.

"Meeting other mothers with similar problems taught us that we were not alone. The rehabilitation program at the March of Dimes Birth Defects Centers in New York City

(which are located at Albert Einstein College of Medicine; Columbia University-Presbyterian Medical Center; Hospital for Special Surgery; Jewish Hospital and Medical Center of Brooklyn; The New York Hospital-Cornell Medical College, and New York University Medical Center, and are supported by the New York Chapter of the National Foundation-March of Dimes) enable us to observe how these children progress. It also helps us accept our daily problems. However, through these contacts we have the opportunity to put aside our personal preoccupation with our own children's problems by sharing our experiences with others.

"We realize that there is no complete solution for our complex and fluctuating feelings. Yet, these experiences have helped us find means of living more normally in an abnormal situation.

"After taking our children home, friends and relatives helped by allowing us to talk realistically about the situation. Some of us found that friends were often more helpful than relatives because they were less involved emotionally, and therefore, more objective.

"We welcomed comfort from those who showed compassion without displays of pity. Others offered sympathy in a way that increased our own sense of guilt, shame, or self-pity. We came to realize that they meant well but lacked understanding of our innermost feelings. This realization helped us to overcome these disturbing emotions.

"Often uninformed friends and relatives offered advice which only added to confusion and frustration. We found that this was best handled by thanking them for their consideration, but by following only the suggestions of qualified persons.

"As time passed, the curiosity of outsiders was more easily handled. Each mother must decide how much information she wants to give. In some instances, a detailed description of the condition helps others understand the child better. At times it may be better to answer as briefly as possible. We have learned, too, that, as the child matures, explaining the problem bit by bit will help him answer outsiders' questions.

"We found that by talking to other mothers at the Birth Defects Center we were able to see much that we had overlooked.

"Some of the basic things we now know are that we all tended at first to focus on the handicaps of the child, rather than on the child himself. For instance, one mother discovered that the only attention she gave her child was directed towards his carrying out his therapy. She had rarely found time to play games or read to her son. True, therapy for the child must be a part of his daily routine, so that it becomes habitual like brushing his teeth. But 'fun and games' are equally important.

"At first, some of us gave excessive attention to the handicapped child. We have learned from the professionals who work with us that we must fit the child into the family instead of the family to the child. One mother realized the truth of this only after months of concentrating on therapy, while her housework, laundry, and other necessary tasks piled up.

"If the child is part of the family, it is better to teach him to do things for himself. This is as important for a handicapped child as for a normal one. Whenever possible, he must learn to tie his shoes, dress, feed, and entertain himself. This often has to be explained to others, who tend to resent any effort to discipline a disabled child or make normal demands of him. Some of us must ignore oversolicitous outsiders if they cannot understand this explanation.

"To reduce the danger of unthinking pity by others, we first had to eliminate our own. We have learned to focus on our children's abilities instead of their disabilities. We have taught our children that walking with braces and crutches is an accomplishment rather than a limitation . . . .

"We find it helpful to look each day for small accomplishments. This way the child's progress seems faster, and boosts his and our morale. We have come to understand and appreciate the meaning of the old saying, 'The longest journey starts with a single step.'

"In the beginning most of us felt that our husbands' attitudes were different from ours. However, we soon realized that men vary a great deal, in their outlook, just as women do. Whether they see the child's future optimistically or pessimistically seems to depend on personality, home environment, family relationships, and whether the child is a boy or girl. Nevertheless, we did find some general differences between the attitudes of our husbands and ourselves.

"For example, husbands express much more concern for a boy's than a girl's future. They are understandably anxious about how he will make a living.

"Most of our husbands felt insecure in caring for the children, because they were away from home most of the day and were consequently unfamiliar with the handling of a disabled child. However, many fathers seemed much more efficient than the mothers in motivating their children.

"We realize now more than ever before how husbands and wives really depend on each other. When one is depressed or excited, the other often seems able to remain calm and reliant. This kind of balance and stability is important to every child, whether handicapped or not. . . .

"The March of Dimes Birth Defects Center is set up to focus on the total child, not just one aspect of his problem. Upon entering the Center, we meet a team of specialists who continue regularly to follow our children. The medical team consists of a pediatrician, who cares for the child's general health; a neurologist, who checks the neurological functions; a neurosurgeon, who may repair the spine and may insert a shunt—he regularly checks the shunt to insure proper working and also advises regarding any needed revision as the child grows; a urologist, who checks the bladder and kidneys—children are put on medication to prevent bladder infection, and a urine analysis is made at each clinic visit. In certain cases, the mother is taught to Crede (applying pressure with hands or knuckles to the abdomen below the navel), to empty the bladder and to keep infection from starting. Most of our children cry while being Creded, but it does not really hurt; we use various methods of this and in time the children are taught to do it themselves; and last, an orthopedist, who checks the legs for weakness and deformity and prescribes the braces, appliances, and physical therapy.

"There are also others who are involved in the medical program, such as ophthalmologists, and physical therapists. A physical therapist sees many of our children once a week or more and teaches them to use the various daily exercises. The first stage of therapy is to strengthen the muscles of the trunk and arms.

"Orthopedists advocate early bracing for many of our children. One of the reasons for this is that a child who can stand upright sees the world in a new light, rather than from

the disturbed perspective from a wheelchair or bed. In braces the child is taught to stand in parallel bars, and then to stand with crutches. A mirror at one end of the parallel bars lets the child watch each small step as he takes it. A game of kickball is fun for the beginning and encourages him to move forward after the ball. Doing push-ups from a small seat strengthens the arm muscles for crutch use. Letting a child do push-ups in front of his favorite television program is entertainment and exercise at the same time. Gradually, our children learn to walk with crutches, first with assistance and then on their own. After accomplishing this, they are taught how to fall without hurting themselves, and to get up alone.

"There may be occasional setbacks such as after operations or on removal of braces. These may seem to reduce tediously acquired accomplishments and may cause lack of confidence and the return of past fears. But with strong professional help, parental encouragement, and diligent exercise, these can be overcome.

"Social workers are available at the Centers to help us cope with the various social difficulties in having handicapped children.

"Naturally, the early treatment of our children is directed towards enabling them to attend school. Some will be able to attend regular classes in local or parochial schools. Some will attend school in special classrooms for handicapped children. The exact type of school will depend on the local school facilities, on parental attitudes, and on the child's mental and physical ability."

Milestone on the March:

When you realize that only 12 years (at this writing) have passed since The National Foundation took up the fight against birth defects, then you may recognize how many people, hours, dollars, and hopes have done so much for so many.

Actually, the first March of Dimes Defects Center opened only in January, 1960, at Children's Hospital, Columbus, Ohio, and currently there are over 100. Today, there are so many plans, so many accomplishments, that only more people and money can continue to make these all possible.

If you have something to contribute for the work of The National Foundation, then do so. If it is money, fine. If it is

only time, still finer. If it is both, then you are truly blessed to give so much from both directions.

Meanwhile, you must also not forget the other organizations whose work may vary from or supplement that of The National Foundation. When one or more of them helps you and your handicapped child, then you must in some way try to help them, also.

It is only by giving—with an open heart, as well as an open mind and hand—that we can afford to accept all that The National Foundation and these other organizations give to us and our families.

New York City
August 20, 1971

Jessyca Russell Gaver

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